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Ayş
Differential Diagnosis of Anechoic Images in Endobronchial Ultrasound

Endobronşial Ultrasonda Anekoik Görüntülerin Ayırıcı Tanısı

Ester Cuevas¹, Yuliana Pascual-González¹, Nikos Koufos², Antoni Rosell³, Noelia Cubero¹

Abstract

In the present pictorial study, we report on four different cases with Endobronchial Ultrasound anechoic images, taken during a mediastinal lymph node study. Each anechoic image had a nodal coagulative necrosis sign. Although the anechoic images had similar characteristics in all cases, the final pathology report was different, with two cases positive for malignancy, one case showing a benign tumor of thyroid gland origin, and one case with tuberculosis. All patients signed informed consent for investigation with the EBUS technique. The procedure was performed in accordance with the regulations of our hospital, and following the usual technique followed in diagnostic mediastinal pathology studies.

Key words: EBUS-TBNA, lymph nodes, anechoic images, hypoechogenicity, heterogeneity.

Özet

Bu görsel ağırlıklı sunuda, mediastinal lenf bezi çalısmaları sırasında, endobronşial ultrasoundda anekoik görüntü saptanan dört farklı olguyu sunuyoruz. Her bir anekoik görüntü, lenf bezinde koagülatif nekroz işaretle vermektediydi. Tüm olgularda anekoik görüntüler benzer karakteristik özelliklere sahip olmasına rağmen, patoloji rapor sonuçları farklıdır. İki olguda malignite pozitif idi, bir olguda tiroidden orijin alan benign tümör ve bir olguda da tüberküloz saptandı. Tüm hastalardan EBUS işlemi için yazılan onamlar alındı. Mediastinal patolojilerin tanısında gerekli genel incelemleri takiben hastanemiz kurallarına göre işlemler yapıldı.

Anahtar Sözcükler: EBUS, Transbronşyal iğne aspirationsyonu, lenf bezi, anekoik görüntü, hipoekojenite, heterojenite.

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Endobronchial ultrasound (EBUS) allows the visualization of mediastinal and hilar structures with real-time cytological and histological sampling (1). The procedure currently has a place in most scientific society guidelines for the diagnosis of mediastinal lymphadenopathies and lung cancer staging (2). During the procedure, the rapid detection and differentiation of a target lesion between adjacent reactive, nondiagnostic nodes may help in reaching faster and safer results. Accordingly, different ultrasonic patterns have been studied aiming to reach a better distinction between benign and malignant nodes before node sampling, based on such parameters as lymph node size, shape, presence of central necrosis, etc. (3). In this small case series we present different diagnostic scenarios of anechoic lesions detected by EBUS, and compare the image characteristics with the final pathology result.

CASE
Case 1: A 70-year-old male with severe COPD was diagnosed with squamous cell lung carcinoma of the right upper lobe following fine needle aspiration (FNA) under CT guidance. A PET-CT was performed, and several hypermetabolic right lower paratracheal and hilar nodes were detected (stations 4R and 10R). During EBUS-TBNA, the nodes were described as oval in shape, with a 12mm short-axis diameter, ill-defined borders and several heterogeneous, avascular and anechoic areas (Figure 1A). The EBUS-TBNA samples (22G needle) showed a fibrous material with a cloudy appearance, compatible with coagulative necrosis. The final pathology examination revealed squamous cell lung carcinoma with areas of necrosis.

Case 2: A 67-year-old female, ex-smoker was diagnosed with stage IIIA lung adenocarcinoma. During EBUS-TBNA staging, a subcarinal adenopathy was found (PET positive nodes). The detected nodes were oval in shape, 13mm in short-axis diameter, had well-defined borders, were highly heterogeneous and had areas of avascular hypoechogenicity (Figure 1B). The samples obtained (via a 22G needle) contained a stringy, liquefied mucoid material of yellow/red color. The final pathology examination revealed an invasive mucinous lung adenocarcinoma.

Case 3: A 47-year-old female was investigated for a lymphoproliferative syndrome. A chest CT scan was suggestive of a multinodular goiter with the co-presence of an upper paratracheal lymphadenopathy. The FNA sample under CT guidance revealed benign thyroid cells, and an EBUS-TBNA was performed to further investigate the enlarged lymph nodes (station 2L). The largest detected node was round in shape with a 6mm short-axis diameter, had well-defined borders, was heterogeneous, and had avascular and anechoic areas (Figure 1C). The EBUS-TBNA samples (22G needle) contained a non-fibrous bloody material with a greasy appearance. The final pathology examination showed a crazy paving pattern with cracking artifacts, suggestive of benign colloid nodular cells of thyroid gland origin.

Case 4: A 53-year-old female was admitted due to fever while undergoing triple immunosuppressive therapy following a recent heart transplantation. A PET-CT scan was performed showing small sized, hypermetabolic cervical, upper right paratracheal and right hilar nodes. An EBUS-TBNA (22G needle) was performed, and round 2R station nodes of 9.86mm in the short-axis diameter with ill-defined margins, heterogeneous, with hypoechogenic content suggestive of necrosis were detected (Figure 1D). The pathology examination revealed necrotizing granulomas compatible with tuberculosis, which was later confirmed by a Löwenstein-Jensen culture.

DISCUSSION
Endobronchial ultrasound sonographic features that are considered to be predictive of benign disease are small in size, round in shape and with well-defined margins, and present with a central hilar structure and nodal conglomeration. Signs of coagulation necrosis with a negative Doppler indicate the presence of necrotic tissue, and can commonly be found in malignant lymph nodes, as well as in benign diseases, such as tuberculosis (4).

![Figure 1: EBUS image of the anechoic, avascular lymph node, with higher heterogeneity (A), EBUS image of the anechoic, avascular and multi-lobulated lymph node (B), EBUS image of the lymph node with round anechoic areas (C), EBUS image of the lymph node with ill-defined margins, and heterogeneous and hypoechogenic content (D)
Signs of nodal coagulation necrosis are caused by ischemia or hypoxia of the nodal cells, resulting in cell death and gradual digestion of the cellular remnants through the process of heterolysis (apoptosis induced by hydrolytic enzymes from the surrounding cells). The normal architecture of the lymph node can be maintained for several days, despite the central necrosis formation. The presences of anechoic areas observed via endobronchial ultrasound are more often malignant than benign (5).

Benign anechoic images are commonly caused by mediastinal cystic lesions described as homogeneous, with a negative Doppler flow or a heterogeneous background due to deposits of such materials as proteins, calcium oxalate crystals, blood or pus (6).

In conclusion, the sonographic features of lymph nodes can sometimes be a useful diagnostic tool during EBUS-TBNA procedures. Signs of nodal coagulation necrosis can commonly be found in malignant lymph nodes, but cannot be considered as a specific sign, and so further histological sampling and analyses should be considered necessary before making a definitive diagnosis.

CONFLICTS OF INTEREST
None declared.

AUTHOR CONTRIBUTIONS
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Approach to Foreign Body Aspiration in an Infant Using a Cryoprobe

Kriyoprob Kullanarak bir Bebekte Yabancı Cisim Aspirasyonuna Yaklaşım

Mohammad Ashkan Moslehi

Abstract

Foreign body (FB) aspiration is a true medical emergency that occurs due to airway obstruction in which immediate removal is crucial. Rigid bronchoscopy is the preferred method for the removal of foreign bodies lodged in the airways, however studies have found that a flexible bronchoscopy can achieve greater success rates. Recently, although there have been reports of a cryoprobe being used for the removal of FBs in adults, in pediatrics, and especially in infancy, there is little experience about its use, in that tracheobronchial FB aspiration is an infrequently encountered event among neonates and in early infancy. This report highlights the efficacy of using a cryoprobe for a flexible bronchoscopy for the management of a retained FB in a young infant.

Key words: Foreign body aspiration, Bronchoscopic treatment, Cryotherapy, Infant.

Özet

Yabancı cisim aspirasyonu, hava yolu obstrüksiyonuna neden olduğu için acil bir tıbbi durumdur ve bu nedenle, bu tür durumlarda yabancı cisim hemen çıkarılması çok önemlidir. Rijit bronkoskopi, yabancı cisimlerin çıkarılmasında ilk tercih edilen yöntemdir, ancak bazı çalışmalar flexible bronkoskopinin de yüksek başarı oranını sağlayabilmesini göstermiştir. Son zamanlarda, erişkinlerde, yabancı cisimlerin çıkarılması, kriyoprob kullanımla ilgili bazı bildirimler vardır. Fakat pediatrik grupta, özellikle yeni doğan ve infant dönemde, nedenlerin karşılıklı bir durum olduğu için kullanımı konusunda çok az deneyim vardır. Biz de infant dönemde yabancı cisim aspirasyonu olan olgumuzda, flexible bronkoskop ile kriyoprob kullanılmanın da etkinliğini vurgulamak ve bu konudaki deneyimlerine katkıda bulunmak için olgumu sunmayı amaçladık.

Anahtar Sözcükler: Yabancı cisim aspirasyonu, Bronkoskopik tedavi, Kriyoretapi, Infant.

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Foreign body aspiration in childhood is a common and potentially serious problem. A thorough history and a physical examination are of paramount importance in the evaluation of a child with a suspected FB, as this can frequently lead to a diagnosis without the need for further diagnostic workups or imaging. The majority of FB aspirations are observed in children aged 4 years and younger (1). Choking, coughing, stridor, wheezing and vocal changes are the most common indications that a may have aspirated a foreign body. Treatment depends on the time of aspiration, and the location, type and size of the foreign body. A bronchoscopy may be needed to remove the item. There are some clear advantages to use of the RB for the removal of a tracheobronchial FB, however modern endoscopic equipment is available in various sizes and configurations to suit the patient’s age and size. Furthermore, both flexible and rigid endoscopic equipment are currently available.

CASE
A challenging case of 7-month-old female infant was presented with progressively increasing stridor, coughing, cyanotic spells and respiratory distress over the past 2 weeks. The parents provided a history, stating that the problem had started while the patient was playing with her elder sister when she started showing the signs of choking and coughing, and became cyanotic and breathless. AN examination revealed severe respiratory distress, inspiratory stridor, localized wheeze and reduced air entry on the left side. Oxygen saturation was variable, but mostly around 80–85% in room air. A chest X-ray showed a left side whiteout lung with collapse due to a severe obstruction on the left side with a mediastinal shift (Figure 1). This case was rare in that it is unusual for an FB to lodge distally in the left bronchus, especially in such a small infant. An urgent RB was performed in the operation room, but the surgeon was unable to extract the FB as it was lodged distally, out of the range of a rigid bronchoscope, and was embedded within granulation tissue. Moreover, the patient’s condition became worse during the rigid bronchoscopy procedure, so the infant was intubated and transferred to the PICU. The surgeon asked our department to manage the patient, and after talking to the parents, they gave us consent to make a further investigation with a fiberoptic bronchoscopy, and if possible, to remove the FB using the Cryoprobe. The patient was transferred to our bronchoscopy suite and underwent our sedation protocol, in which atropine was administered to decrease trachea-bronchial secretions, an intermittent intravenous injection of Propofol for light sedation, and strict dose monitoring to maintaining the patient’s spontaneous breathing. As a primary investigation, first, a 2.8 mm fiberoptic bronchoscope was passed through the ETT. The bronchoscopy confirmed the presence of an FB in the left main bronchus with inspissated thick mucosal secretions and some granulation tissue. To maintain the oxygen level, the long nasal prongs were placed in the hypopharynx above the vocal cords. After applying suction, and applying an epinephrine solution, the patient was extubated, as the author had to use a 4.3 bronchoscope with a 2 mm working channel for the passage of a thin 2 mm cryoprobe (ERBE Elektromedizin GmbH, Germany). The bronchoscope was placed in the best possible position to access the object. Using the cryoprobe, freezing was applied to the head of the FB for 5 seconds. After creating an ice ball formation, the object was disengaged from the mucosa and was moved proximally into the trachea. In the following stage, through continuous freezing (without releasing the foot pedal), the FB was attached tightly to the tip of the cryoprobe and was fully removed en bloc, along with the flexible bronchoscope. The FB was a nut particle (Figure 2). After cryoextraction, the bronchoscope was again navigated into the involved airway to look for any complications and to remove the secretions. Only thick mucosal secretions that had been trapped behind the obliterated bronchus were noted, and were removed by suction through the working channel.

Figure 1: Chest X-ray showing the left side whiteout lung
Approach to Foreign Body Aspiration in an Infant Using a Cryoprobe | Maslehi et al.

The slight bleeding that occurred was easily managed through the installation of diluted adrenaline (1:10000). Nebulization (Salbutamol 1.5 mg, and adrenaline 1:1000 two ml diluted in 2 ml normal saline) and steroids (IV Dexamethasone 1.5 mg every 8 hourly) were applied postoperatively. The post-operative period was uneventful. A chest X-ray obtained after the procedure showed a fully normal expanded lung on the left side (Figure 3).

DISCUSSION
Retained FBs, especially in the pediatric age group, can be one of the most challenging problems for interventional bronchoscopists, and may result in such serious complications as pneumonia, atelectasis or bronchiectasis (2). The risk associated with these long-term complications is increased as the time from aspiration to diagnosis increases. Remained FBs can be classified into two groups: primary (due to late diagnosis or Silence FB aspiration syndrome) and secondary (due to unsuccessful bronchoscopy) (1). RB is the optimum treatment approach to the management of FBs in all ages, however the retrieval of FBs through the use of an RB can be hampered by several factors, including the type and diameter of the FB, the time elapsed since diagnosis, the place in which the FB is lodged and the extent to which it is embedded by granulation tissue. The age of the child is also very important, as in younger children, the diameter of the airway may be too small to pass an RB and accessories (such as forceps, baskets), which increases the chance of unsuccessful attempts at removal. Recently the use of fiberoptic bronchoscopes for the retrieval of pediatric airway FBs has increased. The smaller size and better navigational properties of flexible bronchoscopes permit the examination of the lower airways with fewer traumas, which is not possible with a rigid bronchoscope (3). There have been many studies reporting on the successful use of various instruments, such as forceps or baskets, for the extraction of foreign bodies in the airway (4,5). Although there have been studies into the cryoextraction of FBs, there have been few case reports and studies examining the safety and efficacy of this method in young infants. In the most recent study conducted in pediatrics, the studied cases were older than 10 months and their weight were more than 10 kg. Zhang et al. concluded that removal of foreign bodies from the airways of children using flexible bronchoscopy CO2 cryotherapy may be considered a safe, easy and effective method (6). To the best of the author’s knowledge, the index case is unique, in that the age and weight of the patient were lower than the cases published in the literature to date. The FB was lodged distally in the left lower lobe bronchus, and more interesting, the extraction with the RB wasn’t successful.

CONCLUSIONS
For the bronchoscopic removal of an FB, RB is still the approach favored by most clinicians, although a variety of flexible techniques and tools are available. The index case highlights the successful use of a CO2 cryoprobe for the extraction of a small FB lodged distally in the left lower lobe bronchus that was fully embedded by granulation tissue during a flexible bronchoscopy.

CONFLICTS OF INTEREST
None declared.
AUTHOR CONTRIBUTIONS
Concept – M.A.M.; Planning and Design - M.A.M.; Supervision - M.A.M.; Funding - M.A.M.; Materials - M.A.M.; Data Collection and/or Processing - M.A.M.; Analysis and/or Interpretation - M.A.M.; Literature Review - M.A.M.; Writing - M.A.M. Critical Review - M.A.M.

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Hyponatremia, Cardiac Tamponade and Carcinoembryonic Antigen in Lung Adenocarcinoma

Akciğer Adenokarsinomunda Hiponatremi, Kardiyak Tamponad ve Karsinoembriyonik Antijen

Didar Şenocak, Tezcan Kaya, Kubilay İşsever, Ensar Özmen

Abstract

Cardiac tamponade and severe hyponatremia are life-threatening and significant clinical findings. Severe hyponatremia, carcinoembryonic antigen (CEA) levels above 1000 ng/mL and cardiac tamponade are rare conditions as initial findings of non-small cell lung cancer. We report a 60-year-old man who presented with a cough, shortness of breath, nausea and loss of weight. The CEA level of the patient was 1041 ng/mL. He also had severe hyponatremia and cardiac tamponade. The patient was diagnosed with metastatic lung adenocarcinoma following an evaluation. The patient underwent chemotherapy treatment, but died 4 months after the cancer diagnosis. Severe hyponatremia, very high levels of CEA, and cardiac tamponade as initial findings of lung adenocarcinoma may be predictors of higher stage disease and poor prognosis.

Key words: Carcinoembryonic antigen, cardiac tamponade, hyponatremia, adenocancer.

Özet

Kardiyak tamponad ve ciddi hiponatremi hayatı tehdit edebilen önemli klinik bulgulardır. Küçük hücreli dış akciğer kanserinin başlangıç bulgularının ciddi hiponatremi, 1000 ng/mL’nin üzerinde karsinoembriyonik antijen (CEA) düzeyi ve kardiyak tamponad olması nadirdir. Bu makalede, öksürük, nefes darlığı, bulantı ve kilo kaybı şikayetleriyle başvuran 60 yaşında bir erkek hasta sunduk. Hastanın CEA değeri 1041 ng/mL idi. Aynı zamanda hastada ciddi hiponatremi ve kardiyak tamponad vardı. Araştırma sonucu metastatik akciğer adenokarsinomu tanısı konuldu. Hastaya kemoterapi verildi ve kanser tanısından 4 ay sonra eksitus oldu. Akciğer adenokarsinominun başlangıç bulgularının ciddi hiponatremi, çok yüksek CEA düzeyi ve kardiyak tamponad olması ileri evre hastalığın ve kötü прогноз göstergesi olabilir.

Anahtar Sözcüklер: Karsinoembriyonik antijen, kardiyak tamponad, hiponatremi, adenokanser.

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Lung cancer is one of the most frequently diagnosed malign diseases all over the world and ranks in first place among the causes of cancer death (1). Local symptoms caused by the primary tumor, symptoms caused by metastases or nonspecific symptoms are usually seen at the time of a lung cancer diagnosis. In a small group of non-small cell lung cancer (NSCLC) hypercalcemia, syndrome of inappropriate antidiuretic hormone (SIADH) secretion and Cushing’s syndrome may be the initial findings of the disease (2). Some tumor markers, such as carcinoembryonic antigen (CEA), may be found to be elevated in lung adenocarcinoma patients. The serum CEA level is reported to be linked with the diagnosis, stage and prognosis of cancer (3). Severe hyponatremia, carcinoembryonic antigen (CEA) levels in excess of 1000, and cardiac tamponade, as initial findings of non-small cell lung cancer, is a very rare condition. We present a case in which all three findings were observed at the same time.

CASE
A 60-year-old male patient presented with a cough, shortness of breath, nausea and loss of weight to the emergency department of our hospital. He had no remarkable medical history or drug use, and had a smoking history of 20 pack years. Upon physical examination, the patient had distant heart sounds with no murmur or rhythmic tachycardia, and respiratory sounds were normal. Blood pressure was 100/60 mm Hg and pulse rate was 125/min. There was no sign of apparent edema or hypovolemia. In the lab tests, hemoglobin was 11.5 g/dl, MCV 83.5 fl, CRP 11 mg/l (0–5), sodium 114 mmol/L (136–146), potassium 3.8 mmol/L, glucose 126 mg/dl, urea 21 mg/dl (17–43) and creatinine 0.5 mg/dl (0.6–1.1). The patient was hospitalized in the internal medicine clinic for severe hyponatremia. Spot urinary sodium was 60 mmol/L, urine osmolality was 432 mOsm/kg, serum sodium was 114 mmol/L and serum osmolality was 231 mOsm/kg. The thyroid hormone parameters and cortisol levels were normal. SIADH was considered as the cause of hyponatremia. A hypertonic sodium solution was given intravenously, and fluid restriction was performed. The patient was evaluated for malign diseases. CEA level was found to be 1041 ng/mL (reference range 0–5). Gastro-duodenoscopy and colonoscopy were normal. There was no pathological finding in an abdominal computed tomography (CT). A thorax CT revealed multiple lymphadenopathies (LAP) in the supraclavicular and left axillary regions (max. diameter was 14x10 mm). There were also lymph node conglomerate in all mediastinal regions and bilateral hilar, and a pericardial effusion with a maximum diameter of 1.5 cm (Figure 1). A bronchoscopy was performed, but there was no sign of a mass lesion. The cytology of the bronchoalveolar lavage fluid was found to be benign. A positron emission tomography-CT (PET-CT) revealed many malignant-looking LAPs in the mediastinum and cervical lymph nodes, with elevated FDG uptake in the paratracheal, aortopulmonary and hilar regions of the mediastinum (max. diameter was 2 cm), while a hypermetabolic tissue lesion was noted in the right hemithorax hilar region that could not be distinguished from the lymph nodes. There were also malignant effusion images in both hemithoraces and in the pericardium (Figure 2). An excisional biopsy of the lymph node was made from the right supraclavicular region. At this point, the patient developed dyspnea, an increase in jugular venous pressure and a peripheral edema. A chest X-ray revealed an increase in the cardiothoracic ratio and water bottle sign (Figure 3). The patient had signs of cardiac tamponade, and an echocardiography revealed a pericardial effusion, 2.1 cm in diameter, in front of the right ventricle that led to signs of cardiac tamponade and diastolic collapse in the right ventricle. An immediate pericardiocentesis was performed by the cardiologist, and 3 liters of hemorrhagic fluid was drained. The cytology of the pericardial fluid was found to be malignant, compatible with the diagnosis of adenocarcinoma. A biopsy of the lymph node revealed a metastasis of a less differentiated lung adenocarcinoma. The patient’s dyspnea regressed and his serum sodium level was stable at around 130 mmol/L. As a result, a follow up of medical oncology was recommended and the patient was discharged. The patient was treated with chemotherapy during the medical oncology follow up, but the disease progressed and the patient died 4 months after the cancer diagnosis.

Figure 1: CT of the chest showing the pericardial effusion with a maximum diameter of 1.5 cm
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DISCUSSION

Lung cancer is the leading cause of cancer death in the world (1). In lung cancer patients, the initial signs of a primary tumor can include chest pain, vena cava superior syndrome, dysphagia, hoarseness, dyspnea, cough and hemoptysis (2). Loss of appetite, fever, fatigue, weight loss and bone pain may also be seen as a result of the systemic effect of the cancer (2). In addition, paraneoplastic syndromes like SIADH, hypercalcemia, clubbing and polycythemia may be observed (2).

Hyponatremia is a dangerous electrolyte imbalance that can cause nausea, vomiting, fatigue, muscle cramps, convulsions, coma and death (4). A serum sodium level below 120 mEq/L is defined as severe hyponatremia, which is a life-threatening condition (4). The underlying cause of hyponatremia is usually SIADH in lung cancer patients (4,5). This paraneoplastic syndrome is usually seen in patients with small-cell lung cancer (2). Severe hyponatremia, as an initial finding of NSCLC, is a rare condition that has been assessed in a few case reports (5). Iyer et al. (5) reported on a patient with NSCLC who had hyponatremia at the level of 126 mEq/L, secondary to SIADH, upon presentation. In their case report, brain metastasis was detected and chemotherapy-radiotherapy was started, and the patient lived for fewer than 2 months. In SIADH due to lung cancer, the treatment of the primary disease with surgery, chemotherapy or radiotherapy is recommended (4). Symptomatic cases with hyponatremia may be treated with intra-venous hypertonic sodium, fluid restriction or a vasopressin receptor antagonist (4). In the present study, the patient had severe hyponatremia, with a serum sodium level of 114 mEq/L. Lung cancer presenting with hyponatremia has been linked to poor prognosis and higher stages of disease (5). The present case supports this, given that the patient survived for only 4 months.

Lung cancer rarely presents with cardiac tamponade as the initial finding (6,7). The other common causes of tamponade are acute myocardial infarction, malignancy, infections, autoimmune disorders or surgery, or can be idiopathic (7). In addition to lung cancer, breast cancer, lymphoma, melanoma, mesothelioma and renal cancers are also reported among the malignant causes of tamponade (6).

Kumar A. et al. (8) reported on a 63-year-old woman who presented with signs of cardiac tamponade. An emergency pericardiocentesis was performed and the pericardial fluid cytology revealed metastatic adenocarcinoma of the lung. The survival of the patient was reported 7 months after the diagnosis of malignancy. Cardiac tamponade causes the fluid in the pericardial cavity to block the right heart filling, and leads to a reduction in cardiac output. Hypotension, tachycardia and increased jugular venous pressure can also occur in the presence of cardiac tamponade (7), and this condition may result in death unless urgent intervention is performed. If clinical tamponade is present, pericardial drainage must be carried out as soon as possible. The case in the present study developed cardiac tamponade, and pericardiocentesis was performed immediately. The pericardial fluid cytology indicated an adenocarcinoma metastasis. Pericardiectomy, percutaneous balloon cardiotomy, the creation of a pleuropericardial window, chemotherapy and radiotherapy are the other choices of treatment for tamponade (7).

Nowadays, tumor markers are often used for tumor staging, for the monitoring of treatment response and for the...
detection of recurrences. CEA is a tumor marker that is produced by tumor cells, and that may also increase in lung adenocarcinoma (9). That said, CEA does not increase in all lung adenocarcinoma patients, and serum levels above 1000 ng/ml are rarely seen (9). In a study of 158 lung adenocarcinoma cases, the mean serum CEA level was reported to be 1.88 ng/mL and the CEA level was found to be high in 28.5% of the patients (10). This study concluded that the follow-up CEA level can be a useful for the detection of early recurrence of lung adenocarcinoma (10). CEA serum level has been reported to be an important prognostic indicator in other studies involving NSCLC patients (3). High levels of serum CEA have been found to be associated with more aggressive biological properties of the tumor, the quantity of tumor cells and higher stages of the disease, as an indicator of poor prognosis (3).

**CONCLUSIONS**

We present here a case with cardiac tamponade, severe hyponatremia and a very high level of serum CEA as the first indication of lung adenocarcinoma. It should be kept in mind that the etiology may be lung cancer in patients presenting with these rare findings. Hyponatremia secondary to SIADH, and very high serum CEA levels are rarely associated with NSCLC. The present case suggests that the coexistence of these three clinical features may point to metastatic disease, a high tumor burden and poor prognosis.

**CONFLICTS OF INTEREST**

None declared.

**AUTHOR CONTRIBUTIONS**

Concept - D.Ş., T.K., K.İ., E.Ö.; Planning and Design - D.Ş., T.K., K.İ., E.Ö.; Supervision - D.Ş., T.K., K.İ., E.Ö.; Funding - D.Ş.; Materials - D.Ş., T.K.; Data Collection and/or Processing - D.Ş., E.Ö.; Analysis and/or Interpretation - D.Ş., T.K.; Literature Review - D.Ş., T.K., K.İ.; Writing - D.Ş., T.K. Critical Review - T.K.

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A Case of Asymptomatic Pulmonary Limited Granulomatosis with Polyangiitis

Dilaver Taş, Saime Ramadan

Abstract

A 50-year-old female patient was admitted to the Obstetrics and Gynecology Department with vaginal bleeding. The patient was diagnosed with uterine myoma, and an operation was scheduled. A chest radiography revealed suspicious bilateral nodules during the preoperative evaluation of the patient. A thoracic computed tomography revealed bilateral nodular lesions in the superior segment of the right lower lobe and the posterobasal segment of the left lower lobe. A core needle biopsy of the right lung revealed a granuloma formation, neutrophil and lymphocyte infiltration, and fibrinoid necrosis in the interstitial and perivascular area. c-ANCA positivity was detected in autoantibody tests. Eyes, ears, nose, mouth and nephrological examinations of the patient revealed no pathology. The patient was diagnosed with Pulmonary Limited Granulomatosis with Polyangiitis, based on the present findings. Treatment with low dose weekly oral methotrexate, prednisone and folic acid was planned. The patient underwent a total abdominal hysterectomy and a bilateral salpingo oophorectomy without complications. The case is presented with a literature review given the asymptomatic status of the patient and the rarity of the disease.

Key words: granulomatosis with polyangiitis, pulmonary, asymptomatic.

Anahtar Sözcükler: granülomatöz polianjitis, akciğer, asemptomatik.
Granülomatöz polianjitis (GPA), sık olarak antinötrofil sitoplazmik antikor pozitifliği saptanan ve nekroz, granülomatöz inflamasyon ve vaskülit gibi inflamatuvar reaksiyon paterni gösteren, otoimmün küçük damar vaskülitidir (1). Önceleri Wegener granülomatozis olarak adlandırılmakla birlikte bugün bu ifade kullanılmamaktadır. GPA, klasik tutulum yanında sınırlı tutulum gösterebilir. Klasik formda üst solunum yolu, akciğerler ve böbreklerden tutulum görülür. Sınırlı formda ise renal tutulum görülmez ve genellikle kadınlarda görülür (2).

Bu yazida preoperatif değerlendirme esnasında tarama yöntemlerinde ve vajinal kanama sırasında başka bir şişikâyet olmayan olgu sunulmaktadır. Hastanın asemptomatik olması ve hastalığın nadir görüldüğü nedeniyle, literatür tartışması eşliğinde sundu.

**OLGU**

Elli yaşında kadın hasta, vajinal kanama nedeniyile Kadın hastalıkları ve Doğum Servisi’ne başvurmuş. Yapılan muayene sonucunda ‘miyoma uteri’ tanısı konmuştur. Operasyon hazırlığı yapılan hasta, çekilen PA Akciğer grafisinde pulmoner nodüler segmentler saptanması üzerine Göğüs Hastahlıkları Servisi’ne konsulte edilmiştir. Vajinal kanama dışında bir şişikâyet olmayan hastanın annamnezinde hepattı B taşıncısı olduğu öğrenildi. AST, ALT ve bilirubin seviyesi normal idi. Kan üreni: 120/70 mmHg, nabız: 76/dk, ates: 36,4, oksijen saturasyonu %98 bulundu. Fizik muayenede solunum sesleri ve kalp sesleri dinlenmektedir.

Bu hasta, 1953’de Fienberg sınırlı GPA terimini ortaya attı ve daha sonra Carrington ve Liebow bu konsepti geliştirdi (3). Sınırlı GPA, klinik bulguların bölgesel olarak geliştiği, hastalığın sınırlı formudur. Sınırlı, üst solunum yolu, akciğer ve böbrek tutulumu göstergi olarak değerlendirilmişti. Ancak diğer organ tutulumları da görülmemiştir. İlk olarak, 1953’de Fienberg sınırlı GPA terimini ortaya attı ve daha sonra Carrington ve Liebow bu konsepti geliştirdi (3). Sınırlı GPA, klinik bulguların bölgesel olarak görülmesi, hastalığın sınırlı for- mumdur. Sınırlı forma, üst solunum yolları, akciğer ve böbreklerden sadece bir veya iki aylık bölgede tutulum görülür.

**TARTIŞMA**

GPA, farklı klinik görünümleri olan multisistemik bir hastalıktır. Nekrotizan granulom, nekrotizan glomerulonefritis ve küçük damar vaskülit hastalığın özelliğidir. Genellikle üst solunum yolları, akciğer ve böbrek tutulumu gösterir. Ancak diğer organ tutulumları da görülmemiştir. İlk olarak, 1953’de, Fienberg sınırlı GPA terimini ortaya attı ve daha sonra Carrington ve Liebow bu konsepti geliştirdi (3). Sınırlı GPA, klinik bulguların bölgesel olarak görülmesi, hastalığın sınırlı formudur. Sınırlı forma, üst solunum yolları, akciğer ve böbreklerden sadece bir veya iki aylık bölgede tutulum görülür.
Olgunumuz asemptomatik olması ilginç bir durum olarak değerlendirildi. Asemptomatik olmasının yanında inflamasyon belirteçleri olan CRP ve sedimantasyon normal sınırlarla idi. Sınırlı GPA yanında miyoma uteri ve hepatit B taşıncılığı öyküsü mevcuttu. Genelde GPA’dan inflamasyon belirteçlerinde yüksekseme saptanan (6,7). Inflamasyon belirteçleri, hastalık aktivitesi, prognoz ve immünsupresif ilaçlara karşı terapotik yanıt değerlendirmenin önemini göstermektedir. Olgunumuzda, c-ANCA titreleri ve ’Birmingham vaskülit aktivite skoru’ yararlı bir test olsa da, özellikle CRP’nin, GPA’yı bir rol oynayabileceği ve/veya pulmoner nodül/kitle mevcut idi. Sınırlı GPA ve/veya yatak odasında nagib olduğu ancak bunun dışında organ tutulumu ve hayatı tehdit eden bir hastalık (glomerülonefrit, pulmoner hemorrhaji, serebral vaskülit, nöropati, orbital psödotümör, gastrentestinal kanama, perikardit, myokardit) olmasının durumunda siklofosfamid yerine tercih edilebilir (16, 17). 

Sonuç olarak; olgunumuz asemptomatik olan GPA’nın asemptomatik olabileceği ve hastaların tanısız kalabileceği ve/veya pulmoner nodül/kitle mevcut olduğu ancak bunun dışında organ tutulumu ve hayatı tehdit eden bir hastalık (glomerülonefrit, pulmoner hemorrhaji, serebral vaskülit, nöropati, orbital psödotümör, gastrentestinal kanama, perikardit, myokardit) olmasının durumunda siklofosfamid yerine tercih edilebilir (16, 17). 

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Concurrence of a Large Parosteal Lipoma and Osteochondroma on the Chest Wall

Göğüs Duvarında Büyük Parosteal Lipom ve Osteokondrom Birlikteliği

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Abstract

Chest wall tumors are very rare, accounting for 3.26–5% of all thoracic neoplasms. Chest wall tumors may originate from superficial or deep soft tissues, and from bone and cartilage structures. Lipoma is the most frequent benign tumor of the soft tissue, and those localized on the chest wall are often well-demarcated and larger than those that are superficial. A lipoma that is in contact with the bone is referred to as a parosteal lipoma. Osteochondroma (OC) is a common benign primary tumor of the bone that generally occurs between the ages of 10 and 30 years. It is often seen in the long bones, and costal localization is rare. We present here the case of a 28 year-old female patient who developed a parosteal lipoma with intercostal extension together with osteochondroma in the neighboring bone.

Key words: Lipoma, osteochondroma, costa.

Özet

Göğüs duvarı tümörleri çok nadirdir ve tüm torasik neoplazmların %3,26 – 5’i oluştururlar. Göğüs duvarı tümörleri yüzeyel veya derin yumuşak dokularдан, kemik ve kıkırdak yapılarından köken alabilir. Lipom en sık görülen benign yumuşak doku tümörüdür. Göğüs duvarındaki lipomlar genellikle yüzeyel olanlara göre daha iyi sınırlı ve geniş olan derin lipomlardır. Lipom eklem ile temas halinde ise parosteal olarak isimlendirilir. Osteokondrom kemik dokunun sık görülen, iyi huylu ve genellikle 10-30 yaşlarında görülen primer tümörüdür. çoğunlukla uzun kemiklerde görülür, costal yerleşimleri nadirdir. Biz interkostal uzanmış bir parosteal lipom ve ilişkide olduğu kemikte osteokondrom olan 28 yaşındaki bayan hastayı sunduğumuz durumda.

Anahtar Sözcükler: Lipom, osteokondrom, costa.

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Chest wall tumors are very rare, accounting for 3.26–5% of all thoracic neoplasms. They may originate from soft tissue, bone or cartilage (1). Benign chest wall tumors are often asymptomatic, and usual present as slow-growing palpable masses (2).

Lipomas originating from mature adipose tissue are the most common of all benign tumors of the soft tissue, and can develop in any part of the body (3,4). They are more prevalent between the ages of 50 and 70 years, and in the obese. Lipomas localized to the chest wall are often well-demarcated and larger than superficial examples (2).

A parosteal lipoma is a rare benign fatty neoplasm with an intimate relationship to the periosteum. The incidence of this form of tumor is 0.3% among all lipomas (5).

Osteochondroma is a common benign primary tumor of the bone that generally occurs between the ages of 10 and 30 years (6). Osteochondromas account for 10–15% of all bone tumors, and more than 30% of benign bone tumors. Such tumors commonly occur in the long bones, but rarely affect the ribs (7).

Concurrences of lipoma and osteochondroma have rarely been reported in literature (8,9). We present a case here with concurrent parosteal lipoma and costal osteochondroma.

CASE
A 28-year-old female patient presented with an approximately 3–4 month history of swelling in left chest wall. A physical examination revealed a palpable, immobile mass lesion measuring 15x13 cm size at the level of 5th to 8th ribs.

A thorax computed tomography revealed a gross mass lesion on the left side with adipose density, starting adjacent to the medial side of scapula, localized between the bone structure and muscle layer on the chest wall, and extending into the thorax through the destruction of a lateral part of the left 7th rib. The lesion’s intrathoracic component had lobulated contours and few thin septa. At its center, the lesion showed an expansible appearance towards the neighboring rib, and contained a bone component approximately 21x30 mm in size that was considered to be reactional (Figure 1A). Thorax magnetic thoracic imaging revealed a cystic heterogeneous lesion of approximately 14x8 cm in size with lobulated contours on the left lateral chest wall, causing the destruction and expansion of the neighboring lateral arches, extending inside the thorax, and causing an elevation of muscle and the subcutaneous layers. The lesion showed no contrast uptake upon the intravenous administration of a contrast material.

Based on these findings, an operation was planned, and a left posterolateral thoracotomy was subsequently performed, for which the muscle layers were separated to allow access to the lesion. The lesion was observed to be a limp, encapsulated mass measuring approximately 15x13x8 cm. The lesion was carefully pushed aside to allow entry to the thorax via the 6th intercostal space. The lesion was fixed to, and had invaded, the 7th rib, extending nearly 4 cm toward the thoracic space. No relationship with the lung parenchyma was observed. Upon separation from the surrounding tissues, the mass lesion was excised via a resection of the rib, including a safe margin of 4 cm at both the anterior and posterior of the rib (Figure 1B). The defect was closed successfully by the muscles and intercostal sutures; no prosthesis was needed for the closure of the thoracotomy.

Pathology reported a parosteal lipoma and osteochondroma of the invaded rib. The surgical border at the 7th rib remained tumor free. The patient was followed up for 3 years without complications.

Figure 1: CT scan of intra and extra thoracic lipoma (A), Peroperative view of the intra and extrathoracic lipoma (B)

Figure 2: Osteochondroma on the right side, and overlying mature lipocytes that are compatible with lipoma covering the tissue on the left side (H&E Staining, X40)
DISCUSSION

Chest wall tumors constitute 2% of all bone and soft tissue tumors (10). Benign chest wall tumors often present as slow-growing palpable masses (3). Şahin et al. (10) reported that 59.2% of patients with such tumors presented with swelling to the chest wall, 27.2% had chest pain (involving the anterior thoracic wall) and 26.2% had lateral chest pain (involving the lateral thoracic wall). Chest wall tumors may originate from superficial or deep soft tissues, or from bone and cartilage structures (1). Chondroma, lipoma and fibroma are the most common of these tumors (10).

Lipomas are the most common of all the soft tissue tumors (3). They originate from mature adipose tissue; they often grow slowly, and are encapsulated and localized superficially or deeply; and they generally require no treatment until they reach a large size (2-4). The case in the present study had a mass measuring approximately 15 cm. Lipomas are generally observed between the ages of 50 and 70, and often among the obese. The case in the present study was 28 years old, and was deemed overweight based on the body mass index (BMI: 29 kg/m²).

Lipomas of the chest wall are generally well demarcated than the more superficial ones, and are deeply located (2). Some lesions may have hourglass appearance with intrathoracic and extrathoracic components. CT and MRI can reveal regularly bordered mass lesions (3). The lipoma identified in the present case had extrathoracic and intrathoracic extensions, and had regular borders according to imaging studies.

Lipomas that are in contact with the periosteum are referred to as parosteal lipoma (11,12). Intraosseous and parosteal lipomas are rare, accounting for only around 0.3% of all lipomas (11). Lipomas can be calcified or ossified. Parosteal lipomas are often associated with cortical hyperostosis, bowing deformity and pressure erosion of the underlying bone (12). In the case described in the present study, part of the 7th rib had been destroyed by the lipoma, which then extended into the thorax. More than half of all parosteal lipomas occur after the age of 40. Such lesions often have a hard consistency and are fixed to the bone (12). The case in the present study was 28 years old, and while her lesion was deeply located, it could be palpated as slightly limp and immobile mass. Imaging studies revealed a parosteal lipoma fixed to the 7th rib.

Osteochondroma is a common benign primary tumor affecting bone tissue (6). Its prevalence among the general population is 3%. It constitutes 10–15% of all bone tumors and more than 30% of all benign bone tumors (7). It occurs frequently in the 1st to 3rd decades as either single or multiple lesions (6). In an osteochondroma case series of 11 patients, Oruç et al. (7) reported that eight patients in the study were male and three were female; the mean patient age was 27.4 years, and osteochondroma was localized to more than one rib in seven patients, a single rib in three patients, and the scapula in one patient. Consistent with the literature, the case presented here was in second decade of life, and had a single lesion.

The first case of osteochondroma of the rib ever to be reported was described in 1975 by Twersky et al. (6). Osteochondroma is rarely observed outside the long bones (7). It is reported to originate from perichondral defects at the border of the growth plate, and is primarily observed in the cartilaginous areas of the long bones (6), and may rarely be localized to the ribs. In general, 2% of all osteochondromas are localized to the ribs (1,6,7). Osteochondromas of the rib often originate from the osteochondral junction or the vertebral end (1,2,7). The lesion in the present case, however, originated neither from the osteochondral junction nor the vertebral end, but, interestingly, from the lateral part of the rib.

Clinically, osteochondromas are often painless. Symptomatic cases may present with neurological symptoms due to the compression of the neighboring nerves along with the palpable mass (6,7). Costal osteochondroma may present with chest pain, pneumothorax, thoracic outlet syndrome, empyema, hemothorax or laceration of the diaphragm (7). The case in the present complained only of a painless swelling caused by the lipoma.

In a chest X-ray, osteochondroma of the rib may appear as a mass associated with the normal bone (6,7). Such lesions may sometimes confused with pulmonary lesions on an X-ray, and so the localization of the lesion may not be revealed (1,7). Lesion localization, association with the neighboring tissues and morphology can be clearly identified with thorax CT and MRI (2,6,7), and such imaging may provide information about the vascularity of the lesion (2). In the present case, CT and MRI revealed an expansile appearance at the left 7th rib.

Complications associated with costal osteochondroma include pathological fracture, osseous deformity, vascular injury, neural compression and malignant transformation (2), although malignant transformation is rare, with a 1% risk in solitary osteochondroma cases and a 10% risk in multiple cases (1,2). Marcove et al. (13) concluded that
CONFLICTS OF INTEREST

None declared.

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A Case of Granular Cell Tumor of the Mediastinum Treated by VATS and A Review of Literature*  

VATS ile Tedavi Edilen Mediastinal Granüler Hücreli Tümor Olgusu ve Literatür İncelemesi  

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Abstract

Granular cell tumors (GCT) of mediastinal origin are extremely rare. To the best of our knowledge there have been a total of 21 cases reported since the first case was reported by Harrer in 1972. Both benign and malignant forms of GCTs have been reported, and some criteria for malignant forms have been described. Preoperative diagnoses are challenging due to the rareness of the condition. Histopathological studies should include immunohistochemistry. Given the rareness of such tumors in the mediastinum, the findings and the diagnostic yield of PET-CT are still unclear. Herein we present the case of a 50-year-old woman suffering from back pain whose diagnosis was based on a thorax computed tomography followed by PET-CT, and who was treated with a video-assisted thoracic surgical excision, with a final pathologic diagnosis of a “granular cell tumor” of the posterior mediastinum.

Key words: Granular cell tumor, mediastinal tumor, video-assisted thoracic surgery.

Özet

Mediastinal Granüler Hücreli Tümor (GHT) oldukça nadirdir. İlk kez 1972 yılında Harrer tarafından bildirildiği tarihten beri literatürdeki toplam olgu sayısı 21’dir. GHT’nin hem benign hem de malign formları bildirilmiş, malignite kriterleri de tanımlanmıştır. Olguça nadir bir tümör olması nedeniyle ayrıncı tanıda düşünülmüştür. Histopatolojik incelemeler mutlaka immunhistokimyasal çalışmalıdır. Mediastende nadir görülen bir tümörü olduğundan, tanıda PET/BT’nin etkinliği de henüz belirlenmemiştir. Sunulan olguda, sırt ağrısı ile başvuran 50 yaşında bir kadın hastaya, toraks computed tomoğrafisi ve takiben PET/CT çekilmesi, posterior mediastinal bölgeye sahip lezyona yönelik VATS ile total exision yapılmıştır. Histopatolojik tanı “granüler hücreli tümör” olarak raporlanmıştır.

Anahtar Sözcüklər: Granüler Hücreli Tümor, Mediasṭinal Tümor, Video-yardımlı Göğüs Cerrahisi.

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Granular cell tumors (GCT) were first described by Abrikossoff in 1926 as a benign neoplasm that is usually characterized by the emergence of painless nodules in superficial soft tissues (1,2). The most common site of involvement is the head and neck region, accounting for nearly two-thirds of all cases, although they can occur in almost any anatomical location (3,4). Mediastinal GCT is an exceptional tumor. The patient characteristics of the 21 cases reported in literature, including the present case, are summarized in Tables 1 and 2. The aim in this report is to distinguish the different properties of this extremely rare mediastinal form of GCT from those seen in other parts of the body by summarizing all of the cases reported in literature to date.

CASE

A 50-year-old woman suffering from back pain for nearly one year was admitted to our clinic, with a medical history of thyroidectomy and cholecystectomy operations in recent years. There were no pathological findings upon physical examination. Her thorax computed tomography revealed a 30x18mm mass lesion located at the level of the 6th thoracic spine in the right posterior paravertebral region (Figure 1). This mass lesion had pathologic FDG uptake (SUVmax: 4.11) on PET-CT (Figure 2), and a higher FDG uptake identified in the nasopharyngeal region (SUVmax: 13.19) led to an otolaryngology consultation being made. The biopsy obtained from the nasopharyngeal lesion revealed “reactive follicular hyperplasia”. There were no pathological findings from a fiberoptic bronchoscopic examination. A video-assisted thoracic surgical excision was planned. A VATS exploration revealed a mass lesion with a smooth and regular surface in the posterior mediastinal region associated with the adjacent sympathetic nerve chain. A total excision was performed.

Upon histopathological examination a 3x2, 5x2 cm sized, solid, yellowish-white mass with a smooth surface with focal areas of bleeding was identified. A microscopic examination revealed a benign lesion with no mitosis, necrosis or nuclear polymorphism, but with an invasion of the fibrous capsule at one point. The tumor was positive for S100 protein, CD68, TFE-3, CD56 and synaptophysin, and negative for EMA and chromogranin in immunohistochemistry. The tumor was also focally positive with PAS staining, and the collagen rich capsule was stained significantly with trichrome. The focal Schwannian differentiation present led a diagnosis of GCT (Figure 3).

DISCUSSION

Granular cell tumor (GCT) is an uncommon neoplasm of Schwann-cell origin that tends to be mostly benign and asymptomatic (1,5). It was first considered to be a type of myoblastoma, however immunhistochemical and electron microscopy studies revealed ultrastructural differences from Schwannian cells (6). It has been reported to occur between the fourth and sixth decades of life, and is more common in women (5). Mediastinal GCT is extremely rare. In this report summarizing all 22 cases of mediastinal GCT reported on to date, we found that patients were between 11 and 66 years of age (median age 34.7), with 11 (50%) younger than 30 years of age. Although some reports suggest that mediastinal GCT is twice as common in women, we identified only a slight female dominance (12 females and 9 males, and one case with unreported gender) (Table 1).
GCT can occur almost anywhere in the body, but the most commonly affected site is the head and neck region, and particularly the oral cavity, followed by breast and skin regions (3,6). Malignant GCTs are reported to constitute only 1–3% of all GCTs (2,4). In our review of literature, 17 of the 22 cases were benign, four were malignant (18.2%) and one was undefined. It can thus be concluded that the probability of malignancy is much higher in mediastinal GCTs than GCTs occurring in the other regions of the body.

The first malignant mediastinal GCT was reported by Harrer in 1972, after which, Fanburg-Smith et al. defined the histological criteria for malignant GCT. Mediastinal GCTs featuring at least three of the following criteria (necrosis, spindling, vesicular nuclei with large nucleoli, >2 mitosis per 10 high power fields, high nuclear/cytoplasmic ratio and polymorphism) are defined as malignant, although one must remember that the only real identifier of malignancy in GCT is metastasis (2-4). Clinically, all of the reported four malignant mediastinal GCTs were symptomatic, and three suffered from dyspnea, yet only one malignant mediastinal GCT was reported to have distance metastasis (rib) and another malignant patient developed liver metastasis in the postoperative sixth month (4,7).

Diagnosing mediastinal GCT is a challenging task. Differential diagnoses include lymphoma, thymoma and neurogenic tumors, which are commonly seen in the mediastinum (6). There is no specific imaging study for the identification of mediastinal masses. Generally, the first approach to defining mediastinal masses, generally shows a well-defined

GCTs, summary of the literature

| Case No | Author          | Age | Gender | Symptom          | Pathology | Relationship with sympathetic chain |
|---------|-----------------|-----|--------|------------------|-----------|-----------------------------------|
| 1       | 1972_Harrer     | 59  | F      | dyspnea, wheezing| malign    | Unknown                           |
| 2       | 1975_Rosenbloom | 11  | M      | asymptomatic     | benign    | +                                 |
| 3       | 1987_Abenoza    | 18  | F      | asymptomatic     | benign    | Unknown                           |
| 4       | 1988_Aisner     | 27  | F      | cough, chest pain| benign    | +                                 |
| 5       | 1988_Robinson   | 27  | M      | asymptomatic     | benign    | Unknown                           |
| 6       | 1998_Smith      | 53  | F      | cough            | benign    | Unknown                           |
| 7       | 2003_Makida     | 21  | M      | asymptomatic     | benign    | +                                 |
| 8       | 2005_Angeles    | 43  | F      | asymptomatic     | benign    | Unknown                           |
| 9       | 2005_Bean       | 51  | M      | dysphagia        | benign    | Unknown                           |
| 10      | 2006_Barrenechea| 38  | U      | asymptomatic     | unclassified| Unknown                        |
| 11      | 2006_Yanagawa   | 16  | F      | unknown          | benign    | +                                 |
| 12      | 2007_Segawa     | 36  | F      | hoarsness        | benign    | +                                 |
| 13      | 2010_Ishibasi   | 21  | F      | unknown          | benign    | Unknown                           |
| 14      | 2011_Ponce      | 28  | F      | asymptomatic     | benign    | Unknown                           |
| 15      | 2012_Nakao      | 66  | E      | back pain        | malign    | +                                 |
| 16      | 2013_Luka       | 64  | M      | dyspnea, cough   | malign    | +                                 |
| 17      | 2014_Kim        | 24  | M      | asymptomatic     | benign    | +                                 |
| 18      | 2015_Kusano     | 36  | M      | cough            | benign    | +                                 |
| 19      | 2015_Shikatani  | 19  | F      | ptosis, myosis   | benign    | +                                 |
| 20      | 2016_Winchester | 16  | M      | asymptomatic     | benign    | +                                 |
| 21      | 2018_Ludovic    | 41  | M      | dyspnea, pain, repeated pericardial effusion | malign | Unknown                        |
| 22      | 2019_Demiröz    | 50  | F      | back pain        | benign    | +                                 |
mass lesion in the mediastinum. Some authors have suggested that magnetic resonance imaging (MRI) is more helpful in distinguishing GCT from other neurogenic tumors of the mediastinum, although the enhancement pattern of GCT is different from the typical patterns of neurogenic tumors (1,6). The role of 18F-FDG PET/CT in GCT is not clear. To the best of our knowledge, our case is one of the three mediastinal GCT cases evaluated by PET/CT, one of which was a malignant mediastinal GCT that had a higher FDG uptake (SUVmax: 12) than the other two benign cases (SUVmax: 4.11 and 4.13 respectively). According to the report by D’Hulst et al. (8), metabolic activity on 18F-FDG PET/CT may help differentiate between benign and malignant lesions, besides assessing the extent of the disease.

Minimally invasive biopsy techniques, including CT-guided biopsy and endoscopic ultrasound-guided fine needle aspiration (EUS-FNA), were previously used to diagnose a mediastinal GCT (9); however it is difficult to confirm all cases of mediastinal tumor pathologically prior to surgery (10). As such, only three of the 22 cases (13.6%) were diagnosed prior to a surgical intervention. It is known that these tumors are not sensitive to chemo or radiation therapy, and the best known treatment is consequently total excision, made with clear margins to prevent local recurrence. As GCTs are usually well circumscribed, VATS is generally the approach of choice for the treatment mediastinal GCT (1).

CONCLUSION

Mediastinal GCT is an extremely rare neoplasia. To the best of our knowledge, this is the 22nd case reported in literature to date. Although there is an insufficient number of cases to support exact conclusions about mediastinal GCT, it can be concluded from the present report that mediastinal lesions are different in many ways from GCTs arising in other regions of the body in terms of their tendency be more malign in behavior, and also their occurrence in the younger population. That said, there is no difference in the histopathologic patterns of mediastinal GCTs and other GCTs. Preoperative diagnosis is challenging, and immunohistochemistry and ultrastructural techniques are required for a definite diagnosis. As it is known that these tumors are not sensitive to chemo or radiation therapy, the optimum treatment is total excision, made with clear margins. All GCT patients should be followed up for local recurrences, with malignant GCTs followed up carefully even in the absence of distant metastasis at the time of diagnosis.

Table 2. The patient characteristics of the all 22 mediastinal GCT cases (this case included) reported in literature.

| Symptom                          | Number | Percent (%) |
|----------------------------------|--------|-------------|
| Asymptomatic                     | 9      | 41          |
| Cough or dyspnea                  | 7      | 31,8        |
| Pain                             | 4      | 18,2        |
| Horner’s syndrome                | 1      | 4,5         |
| Repetitive pericardial effusion  | 1      | 4,5         |

| Side                             |        |             |
|----------------------------------|--------|-------------|
| Right                            | 4      | 18,2        |
| Left                             | 7      | 31,8        |
| Unknown                          | 11     | 50          |

| Treatment                        |        |             |
|----------------------------------|--------|-------------|
| Surgery                          | 12     | 54,5        |
| Unresectable                     | 1      | 4,5         |
| Unknown                          | 9      | 41          |

| Behaviour                        |        |             |
|----------------------------------|--------|-------------|
| Benign                           | 17     | 77,3        |
| Malign                           | 4      | 18,2        |
| Unknown                          | 1      | 4,5         |

| Association with sympathetic nerve |        |             |
|------------------------------------|--------|-------------|
| Yes                                | 12     | 54,5        |
| Unknown                            | 10     | 45,5        |

| Immunohistochemistry               |        |             |
|------------------------------------|--------|-------------|
| S100                               | 12     | 10          |
| NSE                                | 5      | 17          |
| CD68                               | 6      | 15          |
| CD56                               | 3      | 18          |
| Unknown                            | -      |             |

| Stain                             |        |             |
|-----------------------------------|--------|-------------|
| PAS                               | 3      | 19          |

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - Ş.M.D., G.F., G.Y., F.D., P.T.; Planning and Design - Ş.M.D., G.F., G.Y., F.D., P.T.; Supervision - Ş.M.D., G.F., G.Y., F.D., P.T.; Funding ; Materials - Ş.M.D., G.Y.; Data Collection and/or Processing - Ş.M.D., G.F., G.Y., F.D., P.T.; Analysis and/or Interpretation - Ş.M.D., G.Y.; Literature Review - Ş.M.D., G.Y.; Writing - Ş.M.D., G.Y.; Critical Review - Ş.M.D., G.F., F.D.
A Case of Granular Cell Tumor of the Mediastinum Treated by VATS and A Review of Literature | Demiröz et al.

YAZAR KATKILARI
Fikir - Ş.M.D., G.F., G.Y., F.D., P.T.; Tasarım ve Dizayn - Ş.M.D., G.F., G.Y., F.D., P.T.; Denetleme - Ş.M.D., G.F., G.Y., F.D., P.T.; Kaynaklar -; Malzemeler - Ş.M.D., G.Y.; Veri Toplama ve/veya İşleme - Ş.M.D., G.F., G.Y., F.D., P.T.; Analiz ve/veya Yorum - Ş.M.D., G.Y.; Literatür Taraması - Ş.M.D., G.Y.; Yazıyı Yazan - Ş.M.D., G.Y.; Eleştirel İnceleme - Ş.M.D., G.F., F.D.

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Göğüs Duvarı Tümörünü Taklit Eden Primer Sternal Tüberküloz: Olgu Sunumu

Primary Sternal Tuberculosis Mimicking an Anterior Chest Wall Tumor: A Case Report

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Özet

Sternum tüberkülozu tüm kemik-eklem tüberküloz olgularının %1-3 oluştur ve oldukça nadir görülmektedir. Tanı konulması, atipik prezantasyon nedeni ile sık sıkla geçikmektedir. Toraks MR, erken dönem ve atipik prezantasyonlarda tanı的关键 olmakta. 23 yaşındaki kadın hasta, altı aydır göğüs ağrı ve göğüs duvarında şişlik şikayeti ile merkezimize başvurdu. Toraks BT ve MR inda 33x28x42 mm büyüklüğünde sternum korpusunu eroze eden kitle görüldü. Sternum rezeksiyon biopsy samplesi alınarak, histopatolojik incelemesi tüberküloz ile uyumlu bulundu. Ulkemizde tüberkülozun sık görüldüğü bölgelerde genç yaş grubunda sternum tüberkülozu göğüs duvarı kitlelerinin ayırıcı tanıda düşünülmelidir. Göğüs duvarı tümörünü taklit eden ve sternal tüberküloz tanısı kaydıguna-bu olgunsundur. 

Anahtar Sözcükler: granülomatöz polianjiti, akciğer, asemptomatik.

Abstract

Sternal osteomyelitis resulting from tuberculosis (TB) is a clinical rarity, occurring in only 1–3% of all cases of osteoarticular TB. Diagnosis is difficult and is often delayed due to atypical presentation and a lack of awareness. Magnetic resonance imaging (MRI) may be useful in the early stages and in atypical presentations. A 23-year-old female admitted with a 6-month history of chest pain and a mass on middle sternum part of her chest. A computerized tomography (CT) of the thorax and MRI showed a 33x28x42 mm soft tissue mass that was eroding the corpus sternum. Deep biopsy samples from lesions were obtained, and pathology revealed multiple granulomatous and necrotic lesions that were consistent with tuberculous osteomyelitis. The possibility of sternal TB should be kept in mind in the differential diagnosis of masses involving the chest wall, particularly in endemic areas. Herein, we report a case in which a sternal mass mimicked a chest wall tumor that was finally diagnosed as primary sternal tuberculosis.

Key words: granulomatosis with polyangiitis, pulmonary, asymptomatic.
Ülkemizde tüberkuloz insidansı 14,2/100.000 oranında olup, olguların %70’inde akciğer tüberkuloz, %30’unda akciğer dışı organ tüberkuloz görülmektedir. Akciğer dışi organ tüberkülözunda semptom ve bulgular tutulan organa göre değişmekle,plevra tüberkulozu erkeklerde, diğer sistemlere ait organ tüberkulozu ise kadınlar arasında daha sık görülmektedir. Vertebra dışi tüberkuloza bağlı kemik ve eklem tüberkulozu erkeklerde, diğer sistemlere ait organ tüberkulozu ise kadınlar daha sık görülmektedir. Vertebra dışi tüberkuloza bağlı kemik ve eklem tüberkulozunda semptom ve bulgular tutulan organa göre değişmekte, plevra tüberkulozu erkeklerde, diğer sistemlere ait organ tüberkulozu ise kadınlar arasında daha sık görülmektedir. Vertebra dışi tüberkuloza bağlı kemik ve eklem tüberkulozunda semptom ve bulgular tutulan organa göre değişmekte, plevra tüberkulozu erkeklerde, diğer sistemlere ait organ tüberkulozu ise kadınlar arasında daha sık görülmektedir. 

OLGU

Yirmi üç yaşında kadın hasta, 6 aydır devam eden göğüs ağrısı ve göğüs duvarında şişlik şikayeti ile merkezimize başvurdu. Hastanın bilinen herhangi bir hastalığı da operasyon öyküsü yoktu. Hastanın bilinen herhangi bir hastalığı da operasyon öyküsü yoktu. Hasta çalışmıyordu, tüberkuloz temas öyküsü yoktu, sigara ve herhangi bir nedenle ilaç kullanmıyordu. Ateş, kabo kaybı, gece terlemesi gibi hastalık bulguları yoktu. Fizik muayenesinde TA: 110/70, mmHg, nabız sayısı 98/dakika, vücut ısısı 36,5 °C idi. Sistem muayenesinde, inspeksiyonda sternum üzerinde ciltten kabırdık, kızarık ve şişlik mevcuttu. Palpeasyonda bu bölge ağrılı idi. Oskültasyonda bilateral akciğer sesleri doğal idi. Tam kan sayımında ve biyokimyasında özellik saptanmadı. HIV, HCV ve HBV için serolojisi negatif idi. PA akciğer grafisinde parankimal infiltrasyon izlenmedi (Şekil 1). Toraks BT’de sternum corpus proksimal kesiminde destrüksiyon oluşturan fusiform konfigürasyonda, yaklaşık 6,5x3 cm boyutlarında osteomiyelit ile uyumlu lezyon alanı izlendi. Hiler ya da mediastinal lenfadenopati görülmedi. Bilateral parankimal alanlarda aktif infiltrasyon bulgusuna rastlanmadı (Şekil 2). Yüzeyel doku ultrasonografisinde, göğüs duvarı anteriyorda 2.-3. kot mesafesinde sağ sternalost biegle komşuluğunda cilt alta yumuşak dokuda lokalize, iç yapisında difüz mikromilimetrik ekojen gösteren lobüle konturu 27x18mm boyutlarında hipoeokik nodüler lezyon saptanıdı. PPD 15 mm idi. Toraks MR ’da sternum üzerinde yumuşak dokuda şişlik ve loküle dansite artış gözlendi (Şekil 3). Göğüs cerrahisi tarafından yapılan sternal resesi aracılığıyla alınan eksciyonel biyopsi histopatolojisinde bazı alanlarda fokal nerek izlenen kronik inflamasyon ile uyumlu granülom yapıları saptanmııdı (Şekil 4). Doku biyopsi L-J besiyeri kültüründe Mycobacterium tuberculosis complex üredi. İlaç direnç testinde izoniasid, etambutol, rifampisin ve streptomisin hassas bulundu. Bu bulgularla hastaya primer kemik (sternal) tüberkulozu tanısı konuldu, izoniasid 300 mg/gün, Rifampisin 600 mg/gün, pirazinamid 2000 mg/gün ve Etambutol 1500mg/gün tedavi başlandı. Tedavinin inisial fazında antitüberkuloz ilaçlara hepatotoksitite gelişmesi nedeni ile tedaviye 15 gün ara verildi. Karaciğer enzimlerinin normal seviyeye düşmesi ile tedaviye tam doz başlandı ve eksik günleri ilave edildi. Takiplerinde karaciğer fonksiyon testleri normal seyretti, tedavisi 1 yıl tamamlandı. Hastanın sternum üzerinde göğüs ağrısının devam etmesi nedeni ile çekilen kontrol toraks BT ve US bulgularında lezyonun tamamen regresye olduğu görüldü (Şekil 5). Ağınının geçirmiş operasyonu ya da miyaljiye bağlı olduğu düşünüldü. Üç yıldır takip edilmekte olup nüks gelmemiştir.

Şekil 1: PA akciğer grafisi.

Şekil 2: Toraks BT’de sternum corpus proksimal kesiminde destrüksiyon olusturan fusiform konfigürasyonda, yaklaşık 6,5x3 cm boyutlarında osteomiyelit ile uyumlu lezyon alanı.
ŞEKIL 3: Toraks MR'da sternum üzerinde yumuşak dokuda şişlik ve lokale dansite artışı.

ŞEKİL 4: Bazı alanlarda lokal nekroz içeren kronik granülamatöz inflamasyon (H&E X400).

ŞEKİL 5: Kontrol tomografide lezyonlarda regresyon.

**TARTIŞMA**

Tüberküloza bağlı sternal osteomyelit oldukça nadir görülmekte, primer tüberkülozun hematojen veya lenfojen yayımı sonucu, latent odağın reaktivasyonu veya komşu mediastinal lenf nodundan direkt invazyon ile gelişmektedir (3). Osteoartiküler tüberküloz tüm tüberküloz olgularının %2’sini, ekstrapulmoner tüberküloz olgularının %10’unu oluşturmaktadır (4). Ayrıca pediyatrik yaş grubunda BCG aşısı sonrası geliştiği de bildirilmektedir. Sternal osteomyelit genellikle göğüs duvarında travma, mediasinit, subklaviyen ven infeksiyonu ve sternotomi gibi nedenlere bağlı sekonderde oluşabilmektedir. En sık saptanan infeksiyon etkeni Staf. aeurusdur (5). Fulminan tablo ile seyreden infeksiyona bağlı osteomyelitten farklı olarak, tüberküloza bağlı osteomyelitte klinik bulgular sternum üzerinde ağrı şiddetini şeklinde olup sistemik bulgular daha az sıklıkta görülmektedir. Genellikle genç erişkinlerde görülüyebilen, sternal tüberkülozu olan 32 olgunun incelemesi bir makalede, olguların %76’sı erkek, yaş ortalaması 37 (18-74) olarak bulunmuştur (4). Küresel tüberküloz 2016 raporunda, Türkiye’de akciğer dışı organ tüberkülozunun kadınlarda daha sık olduğu, yaş ile akciğer dışı tüberküloz oranının arttığı bildirilmektedir (1).

Olgumuz kadın olup ülkemizdeki akciğer dışı organ tutulumundaki cinsiyet farklılığına uymaktaydı. Yaş ortalaması açısından literatürdeki olgulardan daha genç idi. Sternal tüberküloz olgu serilerinde tanı konulmadan önce ortalaması 6,3 ay bildirilmektedir. Olgumuzda da 6 aydır devam eden sternal tüberkülozu göğüs ağrısı şikayeti mevcuttu. Göğüs radyografisinde kemik destrüksiyonuna yol açan osteolitik lezyonlar, periosital reaksiyon, kalsiyum içeren veya içermeyen yumuşak doku apse formasyonu görülebilir. Bu bulgular osteoartiküler tüberküloza ait patognomik bulgular olduğundan ileri tetik olarak MR veya kemik sintigrafisi gibi inceleme yöntemlerine primer olarak önerilir (6,7). Olgumuzun P-A akciğer grafisinde özellik yoktu. Toraks BT de mediastinal LAM ve parankimde lezyon saptanmadı. BT ve MR incelemede sternum üzerinde destrüksiyon oluşturan osteolitik kitle görüldü. BT kemik tutulumunun değerlendirilmesinde PA grafiye avantaj sağlama amacını beraber mediastinal tutulum ve parankim değerlendirilmede yararlıdır. MR inceleme kemik ve yumuşak dokunun değerlendirilmesinde PA grafinin normal olduğunu hastalığın erken dönemde kemik ilgiçleri keşfedilmedi iyi bir seçeneğe sahiptir. Bir çalışmada olguların %38’inde direkt mikroskopik değerlendirmelerinde, %85’de kültür pozitiflik saptanmış, %67’inde tipik granülom yapısının görülmesi, diğer bir çalışmada ise kemik tüberkülozunda PCR ile M.
tuberculosis saptanmasında sensitivite %85-espesifite %80 olarak bildirilmiştir (4,8). Olgumuzda biyopsi materyalinde L-J kültür pozitifliği saptandı, histopatolojik incelemede granülomların görülmesi ile tanı konuldu. Olgumuzda PCR ile inceleme yapılmadı. İlaç direnç testinde tüm ilaçlara hassas bulundu. Tedavi süresi için ortak bir konsensus olmamakla beraber WHO ve BTS pulmoner ve ekstrapulmoner tüberküloz için aynı süre önerilir (4,8). Operaşyon yerinde akıntı gelişmedi. Kontrol toraks BT ve US da lezyonun regresi olduğu görüldü. Sternal tüberküloza kompleks dekomplikasyonlar gibi osteomyelit, fistül gelişimi, sternumun spontan fraktürü, büyük damarların erozyonu, trakeaya bası, apsesin mediastene, plevraya ve subkutan alana rüptürü görülebilir (4). Tedaviye rağmen iyileşmeyen, komplikasyon gelişen hastalarında cerrahi önerilmektedir. Cerrahi tedavi medikal tedaviyi tamamlayıcı ya da medikal tedaviye yanıt alınamadığı durumlarda, ya da göğüs duvarı kitleye neden olabilecek hastalıklara rıptürü etkilemektedir (4). Yorum - A.B., M.G.O., Veri Toplama ve/veya İşleme – A.B., M.G.O., E.G.C.; Analiz ve/veya Yorum - A.B., M.G.O.; Literatür Taraması - A.B., E.C.; Yazar Yazar - A.B., M.G.O.; Eleştirel Inceleme - A.B., M.G.O.

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ÇIKAR ÇATIŞMASI
Bu makalede herhangi bir çıkar çatışması bildirilmemiştir.

YAZAR KATKILARI
Fikir - A.B., M.G.O., B.A., L.C., M.A.B., E.G.C.; Tasarım ve Dizayn - A.B., M.G.O., B.A., L.C., M.A.B., E.G.C.; Denetleme - A.B., M.G.O., B.A., L.C., M.A.B., E.G.C.; Kaynaklar -; Malzemeler - L.C., M.A.B.; Veri Toplama ve/veya İşleme – A.B., M.G.O., E.G.C.; Analiz ve/veya Yorum - A.B., M.G.O.; Literatür Taraması - A.B., E.C.; Yazar Yazar - A.B., M.G.O.; Eleştirel Inceleme - A.B., M.G.O.
Crohn Hastalığına Bağlı Organize Pnömoni: Olgu Sunumu

Organizing Pneumonia Due To Crohn’s Disease: A Case Report

Melike Yüksel Yavuz, İbrahim Onur Alici, Ceyda Anar, Filiz Güldaval, Melih Büyükşirin

Özet

Ortak embriyojenik köken, otoimmünite, sigara ve kolondan bakteri translokasyonu gibi nedenlerden dolayı akciğer ve bağırak hastalıkları bir arada bulunabilir. İnflamatuar bağırak hastalıklarında (İBH) da Ülseratif kolitli hastalarda, Crohn hastalarında (CH) göre akciğer tutulumu daha sık olmaktadır. Elli üç yaşındaki kadın olgumuz, plöretik ağrı, kuru öksürük ve efor dispnesi ile başvurdu. Antibiyoterapiye yanıt vermemeyen pnömonisi olması üzerine yapılan video yardımcı torakoskopi materyali biyopsi sonucunda organize pnömoni tanısı alındı. Kortikosteroid tedavisi ile klinik ve radyolojik iyileşme gösteren olgumuz, takibinin 18. ayında Crohn hastalığı tanıtıldı. İBH’de hem hastalık ile ilişkili hem de kullanılan ilaç tedavilerine bağlı olarak interstisyel akciğer hastalıklarını görmekteydi. Akciğer patolojileri İBH tansından önce de olabilmektedir. Organize pnömoni, CH’da nadir görülmekte beraber bizim hastamızda olduğu gibi CH tansı sonrasıda da konabilmektedir.

Anahtar Sözcükler: granülomatöz polianjiti, akciğer, asemptomatik.

Abstract

Lung and intestinal diseases may coexist as a result of their common embryonic origin, autoimmunity, smoking and colon translocation. In patients with inflammatory bowel disease (IBD), pulmonary involvement is more common in patients with ulcerative colitis than in patients with Crohn’s disease (CH). A 53-year-old female patient presented with pleuritic pain, dry cough and exertional dyspnea. Antibiotic therapy was initiated, but after the patient did not respond to treatment, a video-assisted thoracoscopy was performed, and a diagnosis of organizing pneumonia was made following the examination of the biopsy material. The radiological and clinical condition of the patient improved with corticosteroid treatment, and a diagnosis of Crohn’s disease was made in the 18th month of follow-up. Interstitial lung diseases can be seen in IBD, related either to the disease itself or to the drugs used. Pulmonary pathologies may also occur prior to a diagnosis of IBD. Although organizing pneumonia is rare in CH, as in our patient, a subsequent diagnosis of CH may be made.

Key words: granulomatosis with polyangiitis, pulmonary, asymptomatic.
Crohn hastalığı (CH) ve ülseratif kolit (ÜK) beraberçe inflamatuar bağırsak hastalıkları (İBH) olarak adlandırılmakta ve bu hastalının yaklaşık % 30’ında bağırsak dışı belirtileri den en az bir tanesi görülmektedir (1). İBH’li olgulara pulmoner kompleksiyonlar İBH’nin ekstraintestinal kompleksiyonları olarak gelistiği gibi saldırılan ilac lara bağlı olarak da gelisembilmektedir. Iskelet, eklem, cilt, göz, vasküler tutulumlara göre daha nadir görülmekle beraber; pulmoner fibrozis, vaskülit, bronşiyolit, akut laerenogratok ve solunum fonksiyon testlerinde bozukluklar İBH’ de bildirilmektedir (1). Pulmoner bulgular ile hastalığın aktivitesi arasında kesin bir ilişki gösterilememiştir. Organize pnömoni (OP) etiyolojisinde, kollajen doku ve vasküler hastalıklar, ilaç reaksiyonları, immünolojik bozukluklar, infeksiyonlar, toksik inhalasyon hasarı ve organ transplantasyonları gibi çeşitli faktörler suçlanmakla birlikte, olguların çoğunda neden saptanamaktadır. Makalede, ilk değerlendirmelerde belirli bir etiyoloji saptanmayan ve kriptojenik OP (KOP) tanısı ile tedavi başlanan bir hasta sunulacaktır. Olguda, izlemcinin 18. ayında CH tanısı alması üzerine OP etyolojisinde yer alan bir hastalığın OP’den çok daha sonra da tanısı alabileceği vurgulamak amacıyla sunduk.

OLGU

Elki üç yaşındaki kadın hasta, yaklaşık 4 haftadır devam eden plöretik tıpte yan ağrısı, non-produktif öksürük ve efort dispnesi mevcuttu. Ev hanımı olan hastanın sigara içme öyküsü ve hipotiroidi dışında ek hastalığı yoktu. Fizik muayenede bilateral inspiratuar krepitan raller mevcuttu. Rutin kan tetkiki sonuçları olağan sınırlardaydı. Solunum fonksiyon testi ve diffüzyon kapasitesi testi istendi fakat hasta uyum sağlayamadı. Posterior-anterior Akciğer grafisinde (PAAG) bilateral alt zonlarda heterojen yaygın, alt loblarda birleşme eğilimi gösteren, hava bronkogramları içeren konsolidasyon alanları izlendi. Solunum fonksiyon testi diagnostik kapatıste testi istendi fakat hasta uyuşu sağlanamadı. Akciğer grafisinde (PAAG) bilateral alt zonlarda heterojen yama tarzi opasite artışı mevcuttu. Hastanın yüksek rezolüsyonlu bilgisayarlı tomografisinde (HRCT), her iki akciğerde yaygın, alt loblarda birleşme eğilimi gösteren, hava bronkogramları içeren konsolidasyon alanları izlendi. Bulguların nedeni olan hastanın sigara içme öyküsü ve hipotiroidi gibi ek hastalıklar yoktu. Fizik muayenede bilateral Inspiratuar krepitan raller mevcuttu. Rutin kan tetkiki testi de olağan sınırlardaydı. Solunum fonksiyon testi ve diffüzyon kapasitesi testi istendi fakat hasta uyum sağlayamadı. Makalede, ilk değerlendirmelerde belirli bir etiyoloji saptanmayan ve kriptojenik OP (KOP) tanısı ile tedavi başlanan bir hasta sunulacaktır. Olguda, izlemcinin 18. ayında CH tanısı alması üzerine OP etyolojisinde yer alan bir hastalığın OP’den çok daha sonra da tanısı alabileceği vurgulamak amacıyla sunduk.

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Organize pnömoni (OP), respiratuvar bronşiyoller, alveoller kanallar ve alveoller içerisinde fibroblastik tıcaçların oluşturulduğu polipoid granülasyon dokusu ile karakterizedir. Mononükleer hücrelerin ve köpük makrofajların değişken derecede interstisyel ve havalı alan infiltrasyonu tipiktir. Etiyolojisinde viral enfeksiyon, aspirasyon pnömonisi ve diğer interstisyel pnömoniler, konnektif doku hastalıkları, toksik gaz inhalasyonu, çeşitli ilaçlar, transfüzyon, maligniteler ve radyoterapi yer alır. Ancak idiopa-tiktir (2). OP genellikle subakut bir seyir gösterir ve tanı öncesinde ortalama semptom süresi 2-6 aydır. En yaygın semptomlar öksürük, nefes darlığı, ateş, balgam, iştahsızlık ve kilo kaybıdır. Fizik muayenede inspiratuar raller duyulabilir.

Sektör kullanılmaktadır. OP olgularının 2/3’ünde tam yanılt, yaklaşık 1/3’ünde progressyon, çok azda 3-6. ayarda spontan düzelmel olur. Önerilen seçkin tedavi metilprednizolon olur. Başlangıç dozu 1-3 ay arasında 1 mg/kg/gün’dür, daha sonra 3 ay süreyle 40 mg/gün’e azaltılır ve 1 yıl boyunca 10-20 mg/gün devam edilir. Tedavi yeterli süre ve miktarda verilmezse relaps gelişebilir. Relaps gelisen olgularda steroid tedavini tekrar başlamanın ise makrolidler (eritromisin), siklosporin ve rituksimab verilebilir (3,4). HRCT’de kalsiyum anormallikleri olabilir (5). Bizim hastamızda da HRCT’de 18. ay sonunda fibrotik değişiklikler mevcuttu. IBH, genetik, çevresel ve immünolojik nedenlerin ortaya çıkardığı, mukozal inflamasyonla sonuçlanan, multifaktöriyel ve heterojen bir grup patoloji olarak kabul edilir (6). IBH tanılı hastalarda akciğer tutulumu nadirdir. Gerçek insidans bilinemekle beraber 1.400 hasta içeren bir serinin retrospektif incelemesinde sadece 3 hasta bronkopulmoner tutulum saptanmıştır (6). IBH olan 52 hastadan oluşan bir seride, CH olanlarda ÜK olanlardan (% 25’e karşı % 6) daha fazla anormal pulmoner fonksiyon testler mevcuttu, ancak IBH’lerin yaklaşık % 50’sinde anormal toraks BT saptanmıştır (7). Yedi haste içeren bir seride ise, hastaların 4’ü CH idi ve mix nötrofılık-eozinofilik infiltratlar, UIP, Langerhans granülomatosisi (mesalamin kullanımı sonrası) ve KOP idi. Diğer üç hasta hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH olgularında klinik iyileşmeye rağmen HRCT’de kalıcı anormallikler olabilir (5). Bizim hastamızda da HRCT’de 18. ay sonunda fibrotik değişiklikler mevcuttu. IBH, genetik, çevresel ve immünolojik nedenlerin ortaya çıkardığı, mukozal inflamasyonla sonuçlanan, multifaktöriyel ve heterojen bir grup patoloji olarak kabul edilir (6). IBH tanılı hastalarda akciğer tutulumu nadirdir. Gerçek insidans bilinemekle beraber 1.400 hasta içeren bir serinin retrospektif incelemesinde sadece 3 hasta bronkopulmoner tutulum saptanmıştır (6). IBH olan 52 hastadan oluşan bir seride, CH olanlarda ÜK olanlardan (% 25’e karşı % 6) daha fazla anormal pulmoner fonksiyon testler mevcuttu, ancak IBH’lerin yaklaşık % 50’sinde anormal toraks BT saptanmıştır (7). Yedi haste içeren bir seride ise, hastaların 4’ü CH idi ve mix nötrofılık-eozinofilik infiltratlar, UIP, Langerhans granülomatosisi (mesalamin kullanım sonrası) ve KOP idi. Diğer üç hasta hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmıştır (8). IBH tanılı hastalarda pulmoner nodül, KOP ve eozinofilik pnömoni-KOP (mesalamin kullanım sonrası) saptanmış (6). Bununla beraber organize pnömoni (%12), interstisyel akciğer hastalığı (%18) diğer sık görülen hastalıklar (11). Solunum fonksiyon testleri (SFT) çalışmalarının çoğun %61. Bununla beraber organize pnömoni (%12), interstisyel akciğer hastalığı (%18) diğer sık görülen hastalıklar (11). Solunum fonksiyon testleri (SFT) çalışmalarının çoğun (10). En sık havayolu hasarı görüldü (10). En sık havayolu hasarı görüldü. Karbonmonoksit diffüzyon kapasitesi testinde (DLCO), rezidüel volüm ve fonksiyonel rezidüel kapasite değerleri ve (9) primitif foregut’ primitif foregut’ sebebiyle iki sistemin de antijenlere duyarlılığı ile sonuçlanır (9).
(12). Asemptomatik hastalardaki başlıca SFT anomaliliği DLCO’da azalmadır. Bu anomalilik hemen hemen CH olgularının yarısında saptanmıştır. Hastamız SFT’lerine uyumsuz olduğu için değerlendirilmedi.

İBH tanısı ile OP arasındaki interval 2 ay-36 yıl arasındadır. Bizim hastamızda da yaklaşık 1 aydır devam eden plöretik tipte yan ağrıısı, non-prodüktif öksürük ve efort dispnesi mevcuttu. İlk tanı anında ve sonraki kontrollerinde sistem sorgulamamızda karnın ağrıısı, ishal, ates, kilo kaybı veya peranal sorunlar olmamasına rağmen OP tanılarından yaklaşık 18 ay sonra CH tanıısı almıştır. Literatürdeki bir başka olgu ise OP tanısından 2 yıl önce CH tanısı almıştır (13). Akciğer grafisinde bilateral alveoler infiltrasyonlar en sık rastlanan radyolojik bulgudur. Bazen gezici geçici periferik alveoler infiltrasyonlar görülür. BT’de periferik ve alt zonları tutan yama tarzi alveoler infiltrasyonlar ve buzulcum nitelikinde yoğunluk artışları artanlı izlenir. Hastamızda antibiyotik tedavisine rağmen gerilememeyen tipik radyolojik tutumlu mevcuttu. UK tanılı bir hastada ise sülfasalazin kullanımı sonrasında akciğer aפקידside OP yerleşimi gösterilmiştir (14). Bronkoalveolar lavaj (BAL) sıvısı lenfositik olup arımış CD4/CD8 saptanır ki bu sarkoidoz için tipik olduğu gibi Crohn hastalarında da görülür (15). Gene granülomatos pulmoner lezyonların görülmesi CH ve sarkoidoz arasında da bir ilişki olduğunu göstermektedir. BAL lenfositozuya pulmoner fonksiyon bozukluk arasındadır bir ilişki yoktur. Ek olarak lenfositik alveolit ile CD4+/CD8+ orani, tedavi ve CH aktivitesi arasında ilişki de bulunulmuştur (16).

Tani için doku incelemesi gerekmemektedir. Bir seride, hastalının % 74’ünde transbronsiyal biyopsis ile % 23’ünde ise cerrahi biyopsis (torakotomi veya VATS) ile tanı konulmuştur (17). Bir diğer seride ise, bu yöntemlerle tanı orani % 31 ve % 63 olarak bildirilmiştir (18). Hastamızda lokal anestezi altında VATS yapılarak tanı konulmuş ve CH komplikasyonu gelişmemiştir. Bu anormallik hemen hemen CH ve sarkoidoz arasında da bir ilişki olduğunu göstermektedir. BAL lenfositozuyla pulmoner fonksiyon bozuklukta bir ilişki yoktur. Ek olarak lenfositik alveolit ile CD4+/CD8+ orani, tedavi ve CH aktivitesi arasında ilişki de bulunulmuştur (16).

OP’nin hastalıktaki aktivitesi ile ilişkisi tam olarak kanıtlanmamıştır. Kolektomi ile OP ilişkisiz saptanmıştır. İBH seyirci kullanılan ilaçlar da OP etiyolojisinde suçlanmaktadır. Sulfasalazin (5-aminosalisilik asit ve sulfopiridin kombinasyonu) ve mesalazin bu ilaçlar arasında sidir. Sulfasalazin ve mesalazine bağlı da OP olguları bildirilmiştir (19). OP diğer interstisyan akciğer hastalıklarına göre daha iyi prognoza sahiptir. Dinneen’ın editöre mektubunda bahsettigi olguda, 10 yıldır CH tanisi vardı ve mesalazinaldan tam yarım alınmış ve mesalazin tedavisine pulmoner semptomlar nüks etmeden tekrar başlanılmıştır (20). Bizim hastamız gastroenteroloji tarafından zaten metilprednisolon tedavisi altında tanı konulduğu için tedavi kesilerek, mesalazin başlanmıştır. Hastamızın takibine devam etmektediyiz. Sonuç olarak, OP tanısı ile izlenen hastalarda altı yata neden zaman içerisinde ortaya çıkabileceğinden, her görülmekte etiyolojiye yönelik çıkarılmasını gerektiği klinisyenler için önem taşımaktadır.

**ÇIKAR ÇATIŞMASI**

Bu makalede herhangi bir çıkar çatışma bildirilmemiştir.

**YAZAR KATKILARI**

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IgG4 Related Disease Imitating Cancer, Autoimmune and Infectious Diseases: A Case Report with Lung Involvement

Neoplastik, Otoimmün ve Enfeksiyöz Hastalıkları Taklit Edebilen IgG4 İlişkili Hastalık: Akciğer Tutulumlu bir Olgu Sunumu

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Abstract

Plasma cell granuloma (PCG) is a rare benign tumor that is difficult to distinguish from malignancy. The terminology associated with PCG is inconsistent, with tumors referred to in literature also as inflammatory pseudotumor, fibrous histiocytoma or fibroxanthoma. Diagnosis: clinical features, serum IgG4 level, radiology and histopathological findings should be evaluated together. We present a case here that is very rare and newly described in literature in which a male patient presented to our clinic with a complaint of hemoptysis, resection due to a lesion on the lung who was subsequently diagnosed with IgG4-related disease.

Key words: IgG4, hemoptysis, fibrous histiocytoma.

Özet

Plazma hücreleri granülomu (PCG), maligniteden ayırt edilmesi güç olan nadir görülen, iyi huylu bir tümördür. PCG ile ilişkili terminoloji ve literatürde tutarsızlık vardır ve bu tümörlerle aynıca enfeksiyöz psödotümör, fibröz histiyositoma veya fibroksantoma da denir. Tanı, klinik özellikler, serum IgG4 düzeyi, radioloji ve histopatolojik bulguların birlikte değerlendirilmesi ile konmaktadır. Çok nadir görülen ve literatürde yeni tanımlanmış, klinikimize hemoptizi şikayetli ile başvuran ve akciğerdeki bir lezon nedeniyle rezeksiyon sonrası IgG4 ile ilişkili hastalık tanısı konulan bu olgumuzu sunduk.

Anahtar Sözcükler: IgG4, hemoptizi, fibröz histiyositoma.

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Immunoglobulin (Ig) G4-related disease (IgG4-RD) was first termed in pancreas and as an autoimmune pancreatitis (1). IgG4-RD is a fibro-inflammatory condition involving tumor growth with multiple lymphocytic leakage agents, including IgG4-positive plasma cells that may contain many regions (2). PCG or inflammatory pseudotumor is a rare lesion that can occur in almost every organ, including the lung, and that is predominantly intraparenchymal. Cardiac and pulmonary involvement, however, is rare (1) PCG abundantly and uniformly infiltrates pulmonary connective tissue, including IgG4 plasma cells, bronchovascular bundles, alveolar interstitium, interlobular septa and pleura. Pleural lesions manifest as a diffuse pleural thickening, accompanied by diffuse sclerosing inflammation and chronic lymphoplasmocytic infiltration, with or without fibrosis. IgG4-RLD lesions mostly develop in peribronchial or perivascular connective tissues, the interlobular septa and the pleura. This distribution is essentially a map of the intrapulmonary lymphatic drainage system, and may be an important step in understanding the pathogenesis. PCG can be interpreted both clinically and radiologically as malignant. Imaging methods such as thorax computed tomography and magnetic resonance imaging are necessary to identify the location and metastasis. It is difficult to distinguish PCG from malignancy and fine needle aspiration, or from histologically frozen sections (2). A complete resection of PCG is necessary to reduce the risk of recurrence. A PCG is usually associated with pathological IgG4 levels, and a high serum IgG4 concentration may help differentiate between PCG and other tumors (3). The presence of a circulating plasmablast may be more sensitive as a marker for elevated IgG4 levels when IgG4-RD is diagnosed, but this has not been investigated in our patient (4-6). In this study, we present a case of PCG.

CASE
A 28-year-old male patient was admitted to our clinic with hemoptysis. The physical examination and medical history were unremarkable. Laboratory parameters were normal. A posterior-anterior chest X-ray showed a minimal increase in density in the right lung middle lobe (Figure 1). A thorax computed tomography (CT) and positron emission tomography (PET-CT) were performed, and a solid mass extending to the pleura in the middle lobe of the right lung measuring 4.5 cm (SUVmax 2.8) was detected (Figure 2). A fiberoptic bronchoscopic examination (FOB) revealed a hemorrhagic appearance in the right upper lobe and middle lobe, although no endobronchial lesion was observed. No signs of malignancy were detected in the bronchial lavage material, aspiration or brush samples. No acidoresistant bacilli (ARB) were observed. A transbronchial fine needle aspiration was performed, but no diagnosis was made. Agglutinin tests for cyst hydatid were negative. Accordingly, we opted for surgery due to the continuation of hemoptysis attacks despite medical treatment. A mass associated with right middle lobe and upper lobe was detected after an exploratory thoracotomy. No frozen, benign / malignant distinction was made. A bilobectomy was performed due to hemoptysis complaints, malignancy suspicion and radical surgery. Histopathological examination: In addition to stromal hyalinization and fibrosis widespread proliferation of plasma cells and occasional lymphoid aggregates were identified. Immunohistochemically, the plasma cells showed positive immunoreactivity with kappa and lambda. Intense IgG and IgG4 positivity was found in the plasma-labeled plasma cells (Figure 3). The present findings were interpreted as IgG4-related disease in the lung. No problems emerged in the following 8-month period.

DISCUSSION
IgG4-RD includes increased serum IgG4 concentrations and pathological findings of lymphoplasmocytic infiltration of IgG4-positive plasma cells with storiform fibrosis, as well as obliterative phlebitis in various organs (7). Diseases associated with IgG4 may involve many organs and may exist as autoimmune pancreatitis, Mikulicz’s disease, Riedel’s thyroiditis, retroperitoneal fibrosis and multifocal fibrosclerosis. Although IgG4 plays an important role in the pathogenesis of the disease, the mechanisms of elevation of IgG4 are not yet understood. T cells are thought to be associated with pathogenesis, having been found in many CD4-T cell inflammation sites in IgG4-related diseases. IgG4-RD of the lung is relatively rare, and may be confined to the lung or may develop simultaneously in other organs, or metachronously (8). The clinical and imaging findings of IgG4-RD are highly variable. While lung parenchymal involvement (mass-like lesion or interstitial lung disease) and mediastinal lymphadenopathy are typical, airway and pleural involvement are rare (9). In fact, several cases of IgG4-RD have been reported in the lung parenchymal interstitium, with or without disease, and in the bilateral pleura (10).
IgG4-related disease (IgG4-RD) may explain a significant subgroup of fibro-inflammatory disorders that are of unknown origin in thoracic medicine, such as inflammatory pseudotumor of the lung (known also as IPT, and as plasma cell granuloma), non-specific interstitial pneumonia, and idiopathic interstitial pneumonia, including cryptogenic promoters’ pneumonia and fibrosis (sclerosing) mediastinitis, which are frequently detected in patients with IgG4-RD. A systematic review of the clinical records of the Mayo Clinic, Rochester, MN revealed 127 cases meeting the current diagnostic criteria for IgG4-RD, and 16 (12.6%) with lung or thoracic involvement (12).

Patients with IgG4-RD have been reported to be at risk of malignancy, suggesting that IgG4-RD is a paraneoplastic syndrome, especially one year after onset. Yamada et al. (13) analyzed 334 patients, in which 67 malignancies were noted in 57 patients, the most frequent of which was lung cancer, in 12 patients. In addition, an analysis of 294 patients with non-small cell lung cancer who underwent a surgical resection found 20 IgG4+ plasma cells per high-power field in 35 patients, of whom six were IgG4 / IgG> 40% (14). These reports suggest a strong association between IgG4-RD and lung cancer, demonstrating the importance of the strict exclusion of lung cancer in the diagnosis of IgG4-RD, and the monitoring of these patients for the development of malignancies.

There are studies suggesting that cancer cells and IgG4-positive plasma cells coexist with the obliterative phlebitis in the same nodule, rather than presenting as an individual complication. The production of IgG4 plasma cells is thought to be a response to unknown antigens. Further research is needed to investigate the pathogenesis of IgG4-RD.

In conclusion, IgG4-related disease contains neoplastic, autoimmune and infectious processes. The clinical, radiological histopathological findings and serum IgG4 level should be evaluated together. The lungs should be kept in mind in the differential diagnosis as a newly defined entity. If possible, a complete resection will be necessary, both for definite treatment and for recurrence.

**CONFLICTS OF INTEREST**
None declared.

**AUTHOR CONTRIBUTIONS**
Concept - O.D., N.O.B., S.C., C.A., İ.Y.; Planning and Design O.D., N.O.B., S.C., C.A., İ.Y.; Supervision - O.D., N.O.B., S.C., C.A., İ.Y.; Funding - O.D., N.O.B., S.C., C.A., İ.Y.; Materials - O.D.; Data Collection and/or Pro-
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