A rare presentation of extragonadal germ cell tumor; massive pericardial effusion with impending tamponade

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ABSTRACT
The clinical manifestations of extragonadal germ cell tumor (EGGCT) depend on its location and are often caused by compression on surrounding structures. Pericardial effusion is absolutely rare as an initial clinical manifestation for this tumor. We report a 31-year-old man presenting to the emergency department with dyspnea. Two-dimensional echocardiography revealed a massive pericardial effusion with impending tamponade. The patient was immediately transferred to the operating room where a cardiac surgeon drained bloody effusion. A mediastinal mass measuring 173×105×105 mm was accidentally noticed during COVID-19 work-up. Fine core needle biopsy of the mass led to the diagnosis of a germ cell tumor, which was treated appropriately. This study shows the importance of proper work-up in pericardial effusion cases with chest CT-scan as an important part of it.

Implication for health policy/practice/research/medical education:
The patient had massive pericardial effusion. A mediastinal mass measuring 173×105×105 mm was accidentally noticed, which led to the diagnosis of a germ cell tumor. This study shows the importance of proper work-up in pericardial effusion cases with chest CT-scan as an important part of it.

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Introduction
The extragonadal germ cell tumor (EGGCT) usually originates from the testis in men. However, a small percentage (<5%) are of extragonadal origin (1, 2).

This tumor is rare and does not primarily involve the testis (1). Its most common extragonadal site is the mediastinum (3, 4). However, primary mediastinal EGGCT is rare and only accounts for 5% of cases with germ cell tumor (5). The symptoms of EGGCT are usually nonspecific and highly depend on the tumor site (6). In cases with anterior mediastinal tumor, the clinical presentation is usually caused by compression on surrounding structures (7). A review of the literature showed that pericardial effusion can rarely be the first manifestation of EGGCT and only few case reports have shown tumoral involvement of pericardium (8-11), which is more common in children and even premature infants (9).

EGGCTs are rare in the anterior mediastinum, manifesting as massive pericardial effusion. However its early diagnosis, immediate treatment, and urgent pericardial drainage are life-saving (8). The etiology of the pericardial effusion can easily be missed in a young or middle-aged person and proper work-up is important in evaluating pericardial effusion.
Case Presentation

A 31-year-old man presented to our hospital, which is a tertiary and teaching center, due to dyspnea and orthopnea. The patient had a history of bipolar disorder. The neck examination was positive for jugular venous distention. The lungs were clear and cardiac examination revealed muffled heart sounds.

The patient was hypotensive with a blood pressure of 90/55 mm Hg, and a heart rate of 110 bpm.

Electrocardiogram revealed sinus tachycardia and low voltage complex. Routine laboratory tests were normal.

The patient was taken to the operating room urgently and underwent subxiphoid drainage in which 600 mL of bloody fluid was aspirated. Pericardial biopsy specimen was sent for histopathological examination and the aspirated pericardial fluid was sent for cytology, biochemistry, culture and cell analysis. A chest tube was inserted into the pericardium.

The day after surgery, the patient underwent a CT scan of the lung to evaluate COVID-19, which revealed a large mass measuring 173×105×102 mm in the anterior mediastinum.

The CT scan reports the presence of various heterogeneous areas, which may be due to degeneration and necrosis in the upper part of mediastinum. The mass lesion was a germ cell tumor. Significant pressure of this mass over the lower part of superior vena cava is seen. A large lymph node (2 mm) is seen adjacent to this mass in the upper anterior mediastinum.

Following this finding, a fine core needle biopsy was performed on the mediastinal lesion, which proved to be compatible with germ cell tumor with extensive necrosis. The result of immunohistochemistry study was CD117; Positive, pan-cytokeratin (panCK); negative, leukocytic common antigen; negative and CD3 was also negative.

The patient was thoroughly examined for the origin of EGGCT and its metastasis. Abdominal and pelvic ultrasound reported no ascites or organomegaly. Bilateral testicular and inguinal ultrasound revealed grade I to II varicocele on the right side with a maximum diameter of 2.8 mm and scattered echogenic microcalculifications (testicular pearls) in the parenchyma of both testes.

A spiral abdominal pelvic multi-slice computed tomography scan with contrast revealed no specific findings, no masses or adenopathy.

A whole-body bone scan with technetium-99m-methylene diphosphonate [(99m)Tc-MDP] revealed a mediastinal mass with no metabolically active bone metastasis. The postoperative course was uneventful and the patient was referred for EGGCT treatment.

Discussion

The mediastinum is the most common site for EGGCT (4) and most of its clinical manifestations occur due to compression on surrounding structures. In the largest study on EGGCT in 600 cases of mediastinal and retroperitoneal masses, the most common manifestations were dyspnea (25%) followed by chest pain (23%) and cough (17%), all were due to the compression. Even a mass weighing 3.5 kg was reported (12) while pericardial tamponade was really rare (6,13).

Tumor rupture can cause pleural or pericardial effusion (14-16) and engage the pericardium. Malignant pericardial effusion is often sporadic and observed in children (8,9,15) and adults (15).

A meta-analysis on the pericardial effusion of 128 patients over 6 years showed that only 12% were initially diagnosed based on pericardial effusion which often accompanied metastasis or pericardial involvement (17), while 87% had a history of malignancy. We introduce a rare case of a 31-year-old man with an initial manifestation of massive pericardial effusion. EGGCT is most prevalent in the third and fourth decades of life (30-41 years) (7, 18). The patient was accidentally diagnosed with EGGCT during the COVID-19 and pulmonary work-up.

Radiological findings can determine the tumor site and origin in the mediastinum, and fine-needle core biopsy is the gold standard for confirming the histological diagnosis (2). Mostly, tumor markers of alpha-fetoprotein lactate dehydrogenase, and beta-HCG indicate germ cell tumors (19).

In our patient, a chest CT-scan detected the tumor, whose diagnosis was confirmed with fine core needle biopsy and tumor markers.

A few cases of EGGCT with the initial manifestation of pericardial effusion (20,8,12,10,16) responded well to chemotherapy and surgical resection.

Conclusion

The clinical manifestations of our patient were hemodynamic compression secondary to pericardial tamponade due to a hidden mass in the anterior mediastinum. This case suggests that these patients should undergo mediastinal examination and CT-scan in addition to cardiac work-up and echocardiography. Examination of the mediastinum can be life-saving and comprises an important part of pericardial effusion evaluation.

Authors’ contribution

MBM was the head of the surgical team who managed this complication. ZAA and HG were a member of the surgical team and helped in data collection. RBM was the anesthesiologist. SAM had a major contribution to the literature search and drafting of the discussion. Hamid Ghaderi drafted the manuscript and provided administrative, technical, and material support. ZAA
edited the final draft. All authors contributed to editing the final draft and approved the manuscript.

Conflicts of interest
The authors declare that they have no competing interests.

Ethical issues
The authors have observed ethical issues including no plagiarism, no data fabrication, no double publication. The patient gave informed consent for the publication of this report.

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