Lymphomas presenting as chest wall tumors

Maligne Lymphome als Thoraxwandtumore

Abstract

Four cases of thoracic lymphoma mimicking chest wall tumors are presented. As resection is not the treatment of first choice in lymphomas, pretherapeutical evaluation of chest wall tumors should include a thoroughly staging and a biopsy for histopathological diagnosis. Chest wall destruction due to an anterior mediastinal mass, or a chest wall tumor associated with mediastinal lymph node enlargement, could be suspicious of thoracic lymphoma. Lymphoma with chest wall involvement mostly turns out to be Hodgkin’s disease or large B-cell lymphoma. Stage and histopathological diagnosis have major impact on treatment and prognosis. Therapy is chemotherapy or chemo-radiation.

Keywords: chest wall tumor, lymphoma, thoracic neoplasms

Zusammenfassung

Wir berichten über vier Fälle thorakaler Lymphome, die klinisch und radiologisch als Thoraxwandtumoren imponierten. Verdächtig auf ein Lymphom sind Tumoren des vorderen Mediastinums mit Destruktion der ventralen Thoraxwand, und Thoraxwandtumoren mit ausgeprägtem mediastinalem Lymphknotenbefall. Histologisch handelt es sich häufig um Hodgkin- oder großzellige B-Zell-Lymphome. Histologie und Stadium sind entscheidend für die korrekte Therapie des Lymphoms, die fast nie in einer Resektion, sondern meist in einer Chemotherapie oder einer kombinierten Chemo-Radiotherapie besteht. Dies unterstreicht die Bedeutung von Histologiegewinnung und Staging bei der prätherapeutischen Abklärung von Thoraxwandtumoren.

Introduction

Primary or metastatic tumors of the chest wall constitute 5% of all thoracic tumors. Resection is the main treatment modality for chest wall malignancy. Only small series are reported in the literature, including malignant soft tissue or bone tumors of the chest wall, invading tumors of bronchogenic or mediastinal origin, local recurrences of breast cancer or metastases of distant malignancies [1], [2], [3]. This selection probably does not reflect the epidemiologic situation.

In 2004, we found four cases of chest wall tumors which turned out to be lymphomas.

Case reports

Case 1

A 32 years old male presented with cough and dyspnea. Thoracic CT showed a homogenous anterior mediastinal mass with vessel and airway compression. Ventraly, the tumor infiltrated and destructed the manubrium (Figure 1). Extrathoracic staging was negative. Biopsies revealed Hodgkin’s lymphoma, nodular sclerosis subtype. A combination chemotherapy plus involved-field radiotherapy was planned.

Case 2

A 39 years old male suffered from chest and shoulder pain for one year. When a swelling at the manubrium arose, corticoids were injected topically. He presented with wound infection and chest wall phlegmona after an incisional biopsy. Thoracic CT revealed a poorly defined tumor destructing the manubrium and sternoclavicular joints, and infiltrating the anterior chest wall and mediastinum. Right paratracheal lymph nodes were enlaged. Extrathoracic staging was negative. Staphylococcus aureus was cultured from wound margins. Infection was managed by surgical debridement and intravenous ampicilline and sulbactame. Histology and immunohistochemistry showed anaplastic large cell lymphoma. After control of infection, combination chemotherapy was started and led to complete remission.
Case 3

A 77 years old male underwent non-conclusive gastroenterologic, urologic and orthopedic examinations for left-sided flank and shoulder pain. Several weeks later, a rapidly growing mass at the 11th and 12th rib was noted. Thoracic and abdominal CT revealed a polycyclic subphrenic chest wall tumor with rib destruction, a second chest wall tumor infiltrating the left deltid muscle and scapula, and mediastinal lymph node enlargement (Figure 2). Biopsies from the rib tumor showed small cell neoplasia. Immunohistochemistry confirmed large B-cell lymphoma. Chemo-immunotherapy led to complete remission.

Case 4

A 69 years old male with history of emphysema, smoking and asbestos exposure presented with weight loss and right-sided anterior chest wall mass. Thoracic MRI confirmed a well defined chest wall tumor with calcifications and destruction of the third and fourth rib. Extrathoracic staging was negative. Biopsies showed a solid, undifferentiated tumor. Immunohistochemistry confirmed large B-cell lymphoma. Therapy was chemo-immunotherapy and radiation.

Discussion

The role of surgery in the management of thoracic lymphoma is to establish the diagnosis by biopsies. If lymphoma is considered, it is almost never a problem to go ahead with the appropriate diagnostic procedure. The typical clinical presentation is a patient aged 25 to 40 years with a mediastinal mass, systemic symptoms as weight loss, fever, and night sweats. For lymph node biopsy mediastinoscopy or anterior mediastinotomy is necessary. Histopathological diagnosis and stage have major impact on treatment protocol and prognosis [4]. Therapy will be chemotherapy or chemoradiation.

Lymphoma represents 5% of all malignancies, and is, compared to chest wall tumors, a common disease. Incidence rates in western countries are 3/100,000 for Hodgkin's lymphoma and 5/100,000 for non-Hodgkin's lymphoma (NHL). The incidence of NHL, especially large B-cell lymphoma, increased during the last two decades [4], [5]. 60% of Hodgkin's lymphomas and 20% of NHL show mediastinal involvement [4]. 20% of all mediastinal masses seen by a thoracic surgeon are lymphomas [4]. Only few reports can be found in the literature dealing with lymphomas invading the chest wall. Surgical data of...
Figure 2: Chest wall involvement (b) and mediastinal disease (a) in large B-cell lymphoma (case 3)
malignant chest wall tumors focus on resected cases and therefore exclude lymphomas [1], [2]. Our case reports show that clinical presentation and imaging of lymphoma may resemble to chest wall tumors, suggesting primary resection. Resection is likely to result in residual tumor, post-operative complications, local recurrence, and systemic progression, and should therefore be avoided. Resection may be an option for chest wall destruction with infection precluding chemotherapy or radiation [6]. Our cases reflect that two subtypes of lymphoma, Hodgkin's disease and large B-cell lymphoma, tend chest wall involvement [4], [7]. For Hodgkin's lymphoma, chest wall involvement is reported in 15% of all stage I-II cases and seems to be an adverse prognostic factor [8]. Chest wall involvement appeared in different stages. It was due to direct invasion from localized disease (case 1 & 2), extralymphatic spread in disseminated disease (case 3), or primary chest wall lymphoma (case 4). In case 1 and 2, involvement of the manubrium led to misinterpretation of primary chest wall tumor. As anterior mediastinal masses are most likely to represent lymphoma and osseous destruction is not uncommon for lymphoma, chest wall involvement is more common in lymphoma than in any other tumors of the anterior mediastinum [7]. Among NHL, mediastinal masses and extranodal involvement are noted most often in large B cell type, as in case 2 and 3. Primary mediastinal large B cell lymphoma is a subtype with unique genetic alterations and aggressive clinical behavior [9]. Case 2 showed extensive chest wall destruction complicated by infection. Case 3 presented a chest wall tumor as first sign of advanced disease. One of the large B cell lymphomas was a solitary chest wall tumor (case 4). This tumor could be a primary malignant lymphoma of the rib. 5% of all extranodal NHL are primary bone lymphomas [10]. Primary B cell lymphomas of ribs were so far only reported anecdotally [11]. There is no consensus about treatment. Combined chemotherapy and radiation is reported to lead to complete remission [10].

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