Case Report

Rare thoracic mass lesion - Myofibroblastoma

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Abstract

Mesenchymal soft tissue masses are uncommon tumours of the chest. Myofibroblastoma is a recently described entity consisting of cells with origin from the myoepithelial cell, mostly seen as benign well-circumscribed neoplasms of the breast tissue. Though usually classified as a benign lesion, rarely it can be multifocal and prone to recurrence. We describe below the case of a 26-year-old female who presented with exertional dyspnoea and evidence of a massive mass lesion in the left hemithorax causing mediastinal displacement to the opposite side. On histological and immunophenotypic analysis, a diagnosis of the very rare thoracic myofibroblastoma was made. The incidence, salient features, pathological differential diagnoses and treatment are reviewed.

Key words: Mesenchymal tumours, Myofibroblastoma, Pulmonary myofibroblastic tumour

Introduction

Mesenchymal soft tissue masses are uncommon but well-described tumours of the chest. Myofibroblastoma, a recently described entity, can rarely present as a chest mass. The term inflammatory myofibroblastic tumour commonly referred to as inflammatory pseudotumour was initially proposed in 1990 in the study of inflammatory lesions of the pulmonary system.[1] Various names such as plasma cell granuloma have been used to describe this entity. Myofibroblastoma has generally been considered a benign tumefaction. Some investigators, however, argue that it may, in fact, be a true sarcoma and prefer the designation inflammatory fibrosarcoma.[2] Currently, it is generally accepted that myofibroblastoma is indeed a true neoplasm with a wide spectrum of biological behaviour, varying from the more frequent benign lesions to the rare tumours that are multifocal and prone to recurrence.[2] Though there are a few reported cases, there is lack of awareness amongst the clinicians and pathologists regarding the presentation and features of this rare entity. We describe below an interesting case of a young female who presented with exertional dyspnoea.

Case Report

A 26-year-old female presented with weight loss and dyspnoea on exertion for 2 months. She had no orthopnoea, palpitations, cough, expectoration, chest pain, haemoptysis or fever. There was no history of pulmonary tuberculosis, collagen vascular disorder or any addiction. Family history and menstrual history were noncontributory.

General physical examination revealed mild pallor and an undernourished appearance. Chest examination showed tracheal deviation to the right, dull left hemithorax with absent breath sounds. Breast and gynaecological examination were noncontributory. All the haematological and biochemical investigations were normal. The sputum was found AFB negative.

Chest X-ray [Figure 1] showed an opaque left hemithorax with mediastinal shift to the right. CECT chest [Figure 2] revealed a large heterogenous extraparenchymal mass-occupying whole of the left hemithorax with complete collapse of the left lung with mediastinal displacement. There was no mediastinal...
lymphadenopathy or rib destruction. The diagnosis of a mesenchymal tumour was suggested. Whole body CT-scan also did not reveal any primary tumour and the breast FNAC was normal. CT guided lung biopsy [Figure 3] showed proliferating pleomorphic spindle cells with vesicular nuclei and pale acidophilic cytoplasm suggestive of actively proliferating myofibroblast with thin collagenous fibres between the cells. Final histopathologic picture was suggestive of a myofibroblastoma. Immunohistochemistry showed vimentin and smooth muscle actin positivity with keratin and S-100 negativity [Figure 4]. A diagnosis of primary myofibroblastoma of the left hemithorax was made. Unfortunately, she expired of progressive respiratory failure before the planned resective surgery. The relatives did not give consent, so an autopsy could not be performed.

Discussion

Myofibroblastomas are rare soft tissue tumours composed of bipolar, uniform, spindle shaped cells arranged in clusters separated by broad bands of collagen. Cells of origin being either fibroblasts, pericytes or smooth muscle cells. Stromal fibroblastic cells with smooth muscle like features are normally seen in the intestinal mucosa, umbilical cord, lymph nodes, and spleen. Electron microscopy of myofibroblasts shows well-developed rough endoplasmic reticulum,
cytoplasmic actin microfilaments, extracellular microtendons, pinocytic vesicles, intermediate and gap junctions.\[5\] Immunophenotypically these tumours express mesenchymal tumour markers like vimentin or desmin and myogenic markers like alpha smooth muscle actin or myosin heavy chain. Most cases of primary neoplasm myofibroblastoma are seen in the male breast tissue as a benign well-circumscribed mass.\[4\]

The lungs are unusual sites for myofibroblastic tumours accounting for just 0.7% of all lung masses.\[6\] These lesions are often discovered incidentally (30%). Patients may present with cough (11%), chest pain (10%), hemoptysis (9%) and dyspnoea (6%). Systemic manifestations, occurring in 30–50% of patients include fever, thrombocytosis, anemia, clubbing and weight loss. These symptoms usually disappear after the tumour is removed.\[1\] Reports of primary pulmonary myofibroblastomas are few. Alam et al\[6\] reported the case of a young female who presented with dermatomyositis and a lung mass that on excision was found to be an inflammatory myofibroblastic tumour. The tumour was excised and the patient survived. Alobeid et al\[8\] reported a series of five cases with a review of 21 previously reported cases showing positivity for actin, desmin and negativity for CD34. Leiomyoma: shows cellular aspirate with eosinophilic cytoplasm containing blunt ended nuclei with immunophenotyping showing positivity for actin, desmin and negativity for CD34. Myoepithelioma: shows keratin and S-100 positivity. So to conclude, myofibroblastomas can rarely present as thoracic mass lesions. These can be diagnosed and differentiated from benign and malignant mesenchymal tumours both by morphology and immunostaining. The highlight of this case is the presentation as a huge mass in early adulthood and the fatal outcome of the ‘benign’ tumour due to respiratory failure.

In the present report the patient presented in early adulthood with slowly progressive symptoms. This late presentation is unique as compared to the more commonly described onset in infancy and childhood. The lack of inflammatory component rules out inflammatory pseudotumour, and the demonstration of vimentin and smooth muscle actin positivity with favourable histology clinches the diagnosis of myofibroblastoma beyond doubt.

Little data is available regarding the management of such tumours. Complete resection of the mass if feasible, is the treatment of choice. Patients with these slow growing tumours have an excellent prognosis if the tumour is completely removed.\[11\] In the absence of such a possibility, radiation therapy,\[2\] immunosuppressive therapy with corticosteroids and nonsteroidal inflammatory agents\[9\] and chemotherapy with or without combined radiation therapy\[10\] can be tried. Although specific management is not clearly defined, the potential use\[10\] of cisplatin, doxorubicin and methotrexate as adjuncts to surgical resection has been proposed, especially for the locally recurrent tumours that are considered as malignant variants.

In the pathological differential diagnoses,\[5\] closest is the malignant fibrous histiocytoma: the most common soft tissue tumour, shows large fibroblastic or myofibroblastic cells with a markedly pleomorphic nucleus or multiple nuclei. The malignant spindle cell tumour: shows lack of cytologic atypia along with absence of haemorrhage, necrosis and mitotic figures. The peripheral neuroectodermal tumours: S-100 positive and negative for actin, desmin and CD34. Leiomyoma: shows cellular aspirate with eosinophilic cytoplasm containing blunt ended nuclei with immunophenotyping showing positivity for actin, desmin and negativity for CD34. Myoepithelioma: shows keratin and S-100 positivity.

So to conclude, myofibroblastomas can rarely present as thoracic mass lesions. These can be diagnosed and differentiated from benign and malignant mesenchymal tumours both by morphology and immunostaining. The highlight of this case is the presentation as a huge mass in early adulthood and the fatal outcome of the ‘benign’ tumour due to respiratory failure.

References


