Intrathoracic Metastatic Low Grade Fibromyxoid Sarcoma

Clinical History:

64 year old man who presented with a progressive shortness of breath over a couple of years. His GP ordered a chest x-ray which showed multiple large round opacities. He also had a long standing enlarged left testicle.

Imaging Findings:

This 64 year old retired aircraft parts engineer presented to his GP with increasing shortness of breath. He is a current smoker of 40 cigarettes a day. His GP ordered a chest x-ray which showed multiple large well circumscribed opacities in both lungs. He also has had a large swollen left testicle for the past eight years. Initially it was thought that the lung lesions could represent a metastatic teratoma, however ultrasound examination and urological review of his testicles did not show any evidence of malignancy.

A CT scan confirmed the presence of multiple oval well defined solid nodular lesions within both side of the lung panenchyma and mediastinum with calcifications.

The patient underwent bronchoscopy which unfortunately did not give a tissue diagnosis. He then had an ultrasound guided biopsy of the lung nodules which was also unsuccessful. He was then referred for a mediastinoscopy. The biopsy of the lesions was successful and material was sent for histology.

Ultrasound of the testicles revealed a large anechoic structure within the left scrotum.

Discussion:

Fibromyxoid sarcomas are very rare tumours often found on the distal extremities. They tend to affect younger patients i.e. <50 years old, the youngest patient reported on was a 4 year old child. They were first described by Evans in 1987 [1]. They often recur after primary surgical excision and frequently metastasise as was the case for this patient. Interestingly no primary sarcomatous lesion was identified for this patient although it may be possible that the original lesion was in the chest wall and at presentation was indistinguishable from the metastases.

A publication in 2000 discussed 11 cases of fibromyxoid sarcoma at a large London teaching hospital. Of these 11 cases the primary lesion was in the lower extremities in four cases, chest wall in three cases, groin in one case, buttock in one case, axilla in one case and retroperitoneum in one case [2]. Of the 11 cases only one patient developed pulmonary metastases.

In this case the histopathology report showed a mixture of patterns with cellular spindle areas, collagenous areas and areas of acellular hyalinisation. These features are likely to represent a fibromyxoid sarcoma.

Immunohistochemistry staining shows nuclear positivity for S100 and cytoplasmic staining with bcl-2. Vimetin is also
strongly positive. An article published in 2007 looked at different histopathological features of 48 low grade fibromyxoid sarcomas. They found that hypercellular areas (16/48), foci of epitheloid cells (13/48), and giant rosettes (6/48) were the most common histological findings. Common immunohistochemical markers present included EMA (41/45), at least focally, CD99 (38/41) and bcl-2 (36/41) while markers were essentially negative for CD34 (2/45), mdm2 (1/41), smooth muscle actin (1/45) and S100 protein (0/46) [3].

Treatment for these patients is centered on surgical resection. The tumours are very unlikely to respond to traditional chemotherapy. Even when successful surgical resection takes place the tumours may recur or metastasise years or even decades later.

In conclusion in a patient with large widespread lesions in the lungs the differential diagnosis should begin with common tumours causing cannon ball metastases but must extend to include soft tissue tumours such as fibromyxoid sarcoma. For this reason a tissue diagnosis is of utmost importance even if the patient has signs of a primary lesion elsewhere.

Differential Diagnosis List: metastatic low grade fibromyxoid sarcoma

Final Diagnosis: metastatic low grade fibromyxoid sarcoma

References:


Description: The chest x-ray ordered by the patients GP showing multiple oval well defined lesions. The lesions were initially thought to represent cannon ball metastases from either a renal, testicular or prostate primary. Origin:
Figure 2

Description: CT slice showing three well circumscribed semi calcified lesions in both lungs. Origin:
Figure 3

Description: CT slice upper chest - four well defined solid nodular lesions with calcifications

Origin:
Description: CT slice (lung window) showing 3 large well circumscribed lesions Origin:
Description: Ultrasound left testicle - no evidence of focal lesion

Origin:
Description: Ultrasound scrotum - evidence of hydrocele

Origin: