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Case Report

Locally Invasive Angiomyolipoma of the Mediastinum

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Abstract

We describe a case of a young woman who developed an epithelial subtype angiomyolipoma of the anterior mediastinum, a rare aggressive form of the tumour not previously reported in the chest. Both radiological and macroscopic appearances of the tumour were abnormal, with an invasive and more diffuse appearing tumour than the classically described localised lesions in the literature.

Keywords: Angiomyolipoma; Mediastinal Tumour; Sarcoma

Case Presentation

We present a case of a 34-year-old lady with an incidental diagnosis of an anterior mediastinal mass during investigations for back pain. She had an Additional past medical history of asthma, breast reduction surgery in 2014, and tonsillectomy in childhood. The mediastinal mass was identified on Computed Tomography (CT) with concerning but non-diagnostic radiological features not consistent with a simple thymoma, lymphoma, teratoma, or thyroid malignancy (Figure 1). Due to the appearance of local invasion, the patient was counselled regarding primary resection due to the high clinical suspicion of malignant potential. Despite this, against medical advice the patient opted instead for radiological surveillance. Excellent performance status and normal pulmonary function tests were noted during work up.

The CT imaging was discussed with several centres throughout the United Kingdom and the consensus opinion was that of an invasive malignancy. A further consultation was undertaken with the patient to present the additional medical opinions and the patient subsequently accepted medical advice and consented to surgical resection. At the time of surgery we adopted a radical approach through a median sternotomy given the radiological concerns of local invasion. The intraoperative appearances mirrored the radiological features of an invasive malignancy, with a partially cystic tumour occupying the anterior mediastinum, invading the pericardium and grossly infiltrating the left phrenic nerve. The mass was removed en bloc along with the underlying pericardium, sacrificing the left phrenic nerve.

Final pathological analysis demonstrated a lesion composed of fibro-fatty tissue with thick walled blood vessels and smooth muscle. The lesion was not well defined and invaded into surrounding thymic tissue and areas of fatty tissue. Despite invasive elements the overall organisation of tissue however favoured an angiomyolipoma.

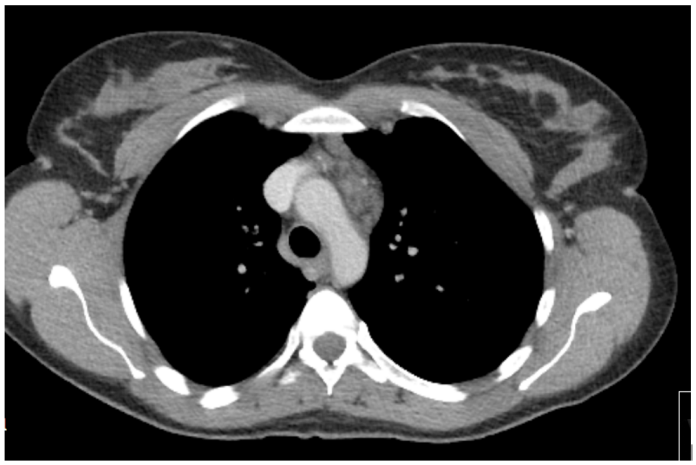

Figure 1

Discussion

Angiomyolipomas are generally benign mixed mesenchymal tumours specifically composed of blood vessels, smooth muscle cells, and fat cells. Whilst most commonly found in the kidney, these tumours can appear sporadically throughout the body particularly in those suffering from tuberous sclerosis [1]. As in our case the majority of angiomyolipomas are asymptomatic on presentation and identified as an incidental finding during investigations for an unrelated condition. Although the reported incidence is increasing in the kidney, mediastinal angiomyolipoma remains exceptionally rare. Angiomyolipomas have additionally been described in the adrenal glands and in the chest wall. To date there have been thirteen cases of mediastinal angiomyolipomas reported in the literature. The authors of these papers generally describe well-defined lesions in keeping with thymomas or schwannomas, depending on their location [1-15]. Direct involvement of adjacent structures is reported in only one other case, with Bai et al describing invasion into the left thoracic cavity from the anterior mediastinum [11]. Candas et al did however report the requirement of phrenic nerve sacrifice to ensure radical resection [12].

Angiomyolipomas arising from the kidney tend to follow a benign course and have a well-circumscribed margin on imaging. However exhibit more alarming features on histological examination with nuclear pleomorphism, microvascular invasion of surrounding blood vessels, and loco-regional lymph node involvement. Despite these features, angiomyolipomas of the kidney still tend to follow a benign clinical course. With few angiomyolipomas reported in the mediastinum, the usual clinical course cannot accurately be described. Notably this is the first cases which describes invasion of mediastinal structures such as the pericardium or phrenic nerves, therefore any discussion surrounding long-term prognosis carries significant uncertainty. The malignant potential and a locally invasive nature of renal based angiomyolipomas has been described in the literature, particularly the epitheloid subtype, but has not been previously observed in mediastinal angiomyolipomas [16]. No lymphatic tissue was identified in the resected tumour which was in keeping with an epitheloid without atypia subtype. This patient was offered surgical excision as her primary intervention however declined and requested radiological follow up. It was only after additional unanimous medical opinions were presented to the patient that she consented to surgery. This reinforces the importance of collegiate working relations between regional centres in managing extremely rare conditions to ensure optimal medical care. In all cases of mediastinal angiomyolipoma described to date, patients have been treated with surgical excision or selective arterial embolisation. Whilst a watch and wait approach is often adopted in benign appearing renal tumours measuring less than 4cm, no papers found in the literature advocate radiological monitoring of mediastinal tumours [17,18].

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