KenKyu

Clinical Case Reports, Research & Trials 2:53-56 (2017)

Case Report

Mediastinal Hemangiopericytoma/Solitary Fibrous Tumor: A rare case

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Received August 30, 2017; **Accepted** November 10, 2017; **Published** November 14, 2017

Abstract

Hemangiopericytoma or Solitary fibrous tumors are rare mesenchymal tumors with very few mediastinal cases reported. Common sites include deep soft tissues of extremities, retroperitoneum, abdomen and trunk.

We report a rare case of a giant mediastinal hemangiopericytoma, in a 56 year old gentleman, presenting with progressive symptoms of productive cough and dyspnea on exertion. Initial discovery on chest X-ray and further workup with a CT scan, led to resection with a median sternotomy. The tumor was large measuring 15cm x 12cm x 9 cm, arising from the anterior mediastinum in the right hemithorax not involving the lung. There was no evidence of malignancy on histopathology. The patient is currently doing well.

The case reports one of the rare vascular and mesenchymal tumors arising from the mediastinum. Given the reported risk of recurrence and increased transformation into malignancy, even years after resection in any hemangiopericytoma, close follow up will be maintained.

Introduction

Hemangiopericytomas a subtype of solitary fibrous tumor (SFT) are rare mesenchymal tumors with very few cases arising from the mediastinum being reported. Solitary fibrous tumor is an uncommon spindle cell neoplasm that was thought to be pleural in origin but extra pleural locations have been increasingly reported. The retro peritoneum, deep soft tissues of extremities, abdomen, trunk, head and neck are the most common [1]. A few cases of mediastinal SFTs have been documented in 1989 by Witkin and Rosai [2] and they are very rare. Hemangiopericytomas were originally described by Stout and Murray in 1942 as a less organoid form of glomus tumor arising from pericytes [3]. These tumors tend to be more common in the skin, subcutaneous tissue, extremities and retroperitoneum. The mediastinum is a very rare location for this disease. Literature review from 2001 revealed only 20 cases in English literature [4]. A PubMed search confirmed very few reported cases of mediastinal occurrence of the tumor. Extra pleural SFTs are are known to remain silent for long periods of time and can attain large sizes. Surgical resection is the treatment of choice. Our patient was found to have a large mediastinal mass demonstrating features of both the above tumor types, which was completely resected without any complications.

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Case report

The patient was a 56 year-old male who was referred to our institution after a chest X-ray was done at his primary physician's office for unremitting symptoms of bronchitis. The Chest X-ray revealed a large mass in the right hemi thorax obscuring the mediastinum and the right heart border. He presented with a 6 month history of exertional dyspnea, pleuritic cough with gray clear sputum and rhinorrhea. He denied any fever/chills. He had been recently started on antibiotics without improvement. He reported a normal chest X-ray done 4 years back. Past medical history was significant for radiation to the thymus gland and left mastoid as a child, history of walking pneumonia, recurrent pleural effusions needed multiple thoracentesis followed by pleurodesis and partial left lower lobectomy for presumed empyema/necrotizing pneumonia. His other medical problems were non-insulin dependent diabetes mellitus, hypertension, obstructive sleep apnea, arthritis. His father had lung cancer and there was history of breast and colon cancer on the mother's side of family.

He smoked pipe weekly since 18 years of age, denied any alcohol or illicit drugs. He worked as a 911 dispatcher.

On examination he was found to have decreased breath sounds on the right compared to left. There was no signs of any shortness of breath or hypoxia. The rest of the exam was within normal limits. A CT Scan of the chest was done which revealed a 10cm mass in the right lung area obstructing the superior vena cava, right up against the chest wall pushing on the mediastinum (Figure.1). CT scan guided biopsy (Figure.2) of the mass showed a spindle cell neoplasm. Histologically there was spindle cell proliferation and densely collagenized stroma with small vessels. Immunohistochemistry demonstrated the cells positive for Vimentin and CD34 but negative for calretinin and cytokeratin. Differential diagnosis was a solitary fibrous tumor as well as other spindle cell neoplasms. The patient underwent median sternotomy and resection of the tumor.

Intraoperatively the mass was located in the anterior mediastinum and had collateral blood supply. It had a vascular attachment to the mediastinal pleura. There was no involvement of the right lung and removal of the mass resulted in complete expansion of the right lung. The gross specimen consisted of a lobulated yellow tan mass measuring 15x12x9 cm (Figure. 3). Sectioning revealed white tan and yellow firm tissue with focal areas of myxoid change without evidence of necrosis (Figure. 4). Histologic review demonstrated a large well circumscribed spindle cell cell neoplasm in association with fibro adipose tissue. The cellularity of the lesion was quite variable with hyper cellular zones alternating with relatively hypo cellular areas with a collagenous stroma (Figure. 5a).

Large zones of ischemic-type necrosis (Figure. 5b) were seen multifocally and a prominent pericytoma like vascular pattern was seen throughout the mass. The lesional cells were arranged as poorly formed short fascicles in some areas and had a haphazard arrangement in hyper cellular zones. The cells throughout the lesion were essentially monomorphic and had irregular oval nuclei with open vesicular chromatin and a small amount of pale to vacuolated cytoplasm. Even in the most hyper cellular areas, mitotic activity

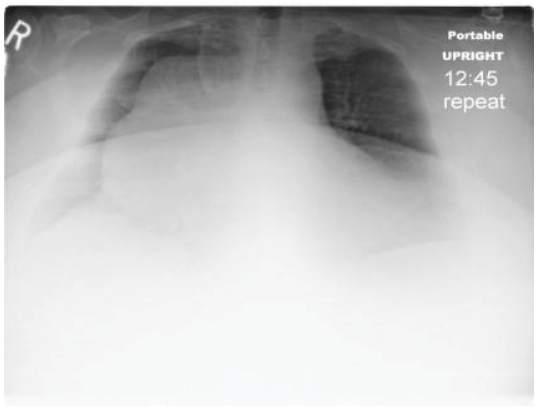

Figure 1: Large mass in right hemi thorax obscuring the right heart Border.

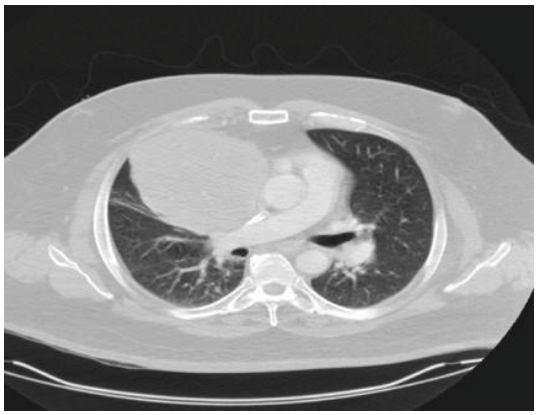

Figure 2: CT scan of the chest with contrast revealed a large 10cm right lung mass silhouetting the mediastinum and obscuring the superior vena cava.

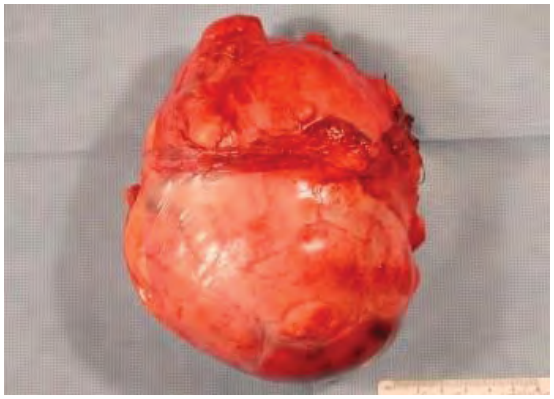

Figure 3: Gross specimen showing well circumscribed yellow, tan lobulated mass.

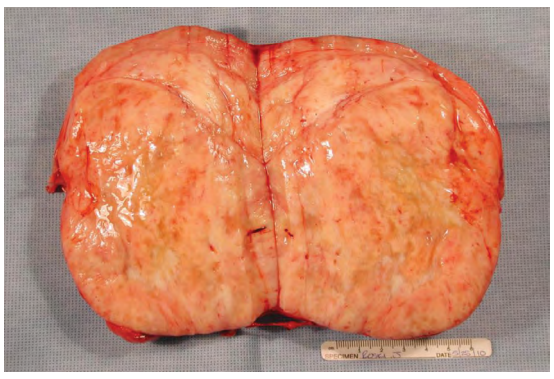

Figure 4: Cut section revealed no evidence of necrosis or hemorrhage.

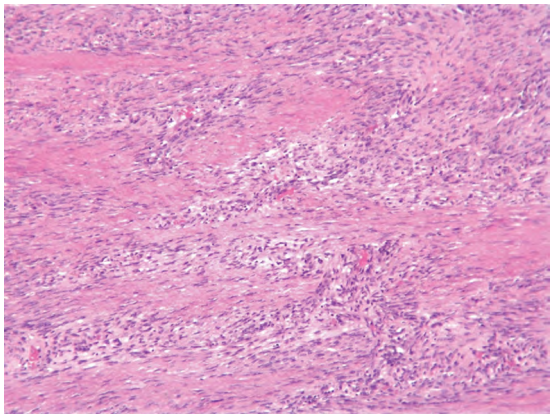

Figure 5a: Variable zones of hyper/hypo cellularity with collagenous stroma and poorly formed fascicles.

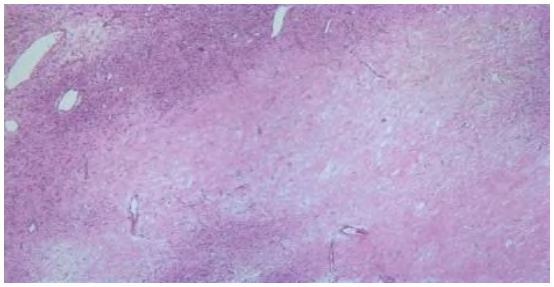

Figure 5b: Large zones of ischemic type necrosis.

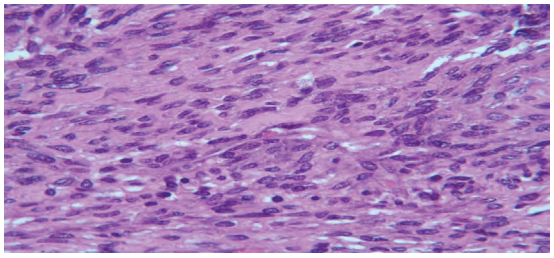

Figure 6: High power view of the neoplastic cells show monomorphic and irregular ovoid nuclei with open vesicular chromatin and a small amount of pale to vacuolated cytoplasm. Overall mitotic activity is low with up to 2 mitoses per 10 high powered fields. (Original Magnification x400).

was quite low with mitoses numbering up to 2/10 high power field (Figure. 6).

Immunohistochemical studies were negative for pan keratin, S 100 protein, SMA and desmin; additional histochemical studies were diffusely positive for BCL-2 and weakly positive for CD 34 in majority of tumor cells.

Discussion

Hemangiopericytoma and solitary fibrous tumors which were initially are classified as separate entities both subtypes of soft tissue sarcoma, falling into the intermediate category of soft tissue sarcomas which rarely metastasize, are now considered together under the spectrum of extra pleural solitary fibrous tumors by the WHO. The mediastinal location is very rare for both these tumors. They are usually thought to arise in the older age group with solitary fibrous tumors occurring predominantly in middle age adults 20-70 years old (mainly 50s) with no sex predilection. They are usually asymptomatic and remain undetected for years, being found incidentally on imaging. But solitary fibrous tumors may present with cough, chest pain and dyspnea [2] as in our patient. It may even present with hypoglycemia. Hemangiopericytoma has been reported to present with massive hemoptysis, spontaneous hemothorax, dysphagia [6-8]. A well-defined round homogenous opacity is usually visualized on a chest X-ray or CT scan [5]. Because of the vascular nature MRI or angiography are thought to be more effective in diagnosis than CT scan [9]. CT scan guided biopsy is not very accurate in establishing a diagnosis [10] and whole tissue is usually needed for diagnosis. Histologic features of malignancy include >4/10 high power field mitotic activity and cellular areas with well-formed fascicular arrangement which were not seen in our patient. These tumors have been shown to follow an indolent but unpredictable course with local recurrence or even distant metastasis. Recurrence has been reported even up to 31 years after surgery in soft tissue fibromas overall and up to 23 years in hemangiopericytoma of the mediastinum [13, 14]. Close follow up has been recommended. Surgical resection is the treatment of choice and patients undergoing complete surgical resection showed 100% survival at 5 years [13]. Preoperative vascular embolization has been recommended for highly vascular hemangiopericytomas [12]. Radiotherapy and chemotherapy have not showed to play a major role and newer therapies are under study for these tumors.

To summarize we encountered one of the rare cases of mediastinal hemangiopericytoma/solitary fibrous tumor which was successfully resected surgically. The patient was doing well after the surgery and we will be closely following up the case.

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