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CASE REPORT

Solitary fibrous tumour of the pleura presenting as a giant intrathoracic mass

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SUMMARY

Solitary fibrous tumours (SFTs) are relatively rare neoplasms thought to originate from the submesothelial connective tissue. SFTs have been described in a variety of sites, including the pleura, orbit, lower respiratory tract, peritoneal cavity and heart. These neoplasms are usually benign, though locally aggressive, and metastatic behaviour has been observed in some cases. We describe a case of a 61-year-old man presenting with weight loss, poor appetite, malaise, worsening dyspnoea on exertion and lower extremity oedema, who was found to have a gigantic—21×21 cm—tumour occupying the entire right hemithorax causing compression and displacement of the mediastinum and liver. Transthoracic CT-guided biopsy revealed SFT of the pleura. The patient underwent preoperative angiography and embolisation of the tumour followed by successful surgical resection via thoracotomy.

BACKGROUND

Solitary fibrous tumours (SFTs) are mesenchymal tumours of fibroblastic origin that can affect virtually any organ. These lesions occur predominantly in middle-aged adults with equal gender distribution.[1] The neoplastic cells are arranged in a patternless architecture with alternating hypocellular and hypercellular areas and a prominent branching vasculature. Most tumours present as well-defined, slow-growing masses, which can be cured by surgery. Surgical resection is the dominant mode of therapy for both benign and malignant SFT of the pleural (SFTP), with a risk of local recurrence possible with the latter even after complete resection. The prognosis for both benign and malignant SFTPs is relatively good. In this article, we report a case of a patient presenting with a massive SFTP occupying the entire right hemithorax and highlight the clinical features, radiographic findings and pathological features of this disease entity. This case highlights the clinical, radiological and pathological features, as well as the role of preoperative angiography and embolisation, in the evaluation and management of massive SFTPs.

CASE PRESENTATION

A 61-year-old man presented to our care with primary complaints of swelling of the bilateral lower extremities and worsening shortness of breath. His symptoms began approximately 4 months prior to admission, with a gradual decrease in appetite and development of progressive weakness and fatigue. The patient had lost up to 10 kg at the time of admission. Approximately 1 month prior to admission, the patient became aware of a palpable mass in his right lower quadrant. He subsequently developed swelling of the right lower extremity, followed by swelling of the left lower extremity. This coupled with worsening dyspnoea on exertion prompted him to seek further evaluation.

He was a non-smoker with a history of heavy alcohol abuse from which he had been abstinent since 1985. He also denied a history of intravenous drug abuse. The patient was unaware of any family history, as he was an orphan. He reported a distant history of an osteosarcoma of the lumbar spine which was resected when he was 19 years old.

At the time of presentation, the patient's vital signs were within normal limits with an oxygen saturation of 98% on room air. He was cachectic in appearance, though generally not ill-appearing. Pulmonary examination was notable for absent breath sounds over the right hemithorax with increased tactile fremitus and increased dullness to percussion. Abdominal examination was notable for a firm right-upper quadrant palpable mass. Lower extremity examination was notable for significant bilateral pitting oedema.

Chest radiograph showed total opacification of the right hemithorax with contralateral mediastinal shift (figure 1). Contrast-enhanced chest CT revealed a 21×21 cm large heterogeneous, non-calcified mass abutting the chest wall with compression and contralateral displacement of the mediastinal structures but without invasion or evidence of significant mediastinal or hilar lymphadenopathy (figure 2). CT of the abdomen and pelvis was remarkable for moderate to severe inferior and lateral displacement of the liver (figure 3). No significant retroperitoneal or intraperitoneal lymphadenopathy was noted. Lung function showed a forced expiratory volume in 1 s and a forced vital capacity of 50% and 48% of predicted, respectively.

The patient underwent percutaneous CT-guided biopsy of the mass. Macroscopically, the specimen consisted of tan-coloured core samples measuring 0.5×0.3×0.3 cm. Histopathological examination demonstrated a tumour composed of haphazardly arranged, bland-appearing spindle cells admixed with thin collagen fibres (Figure 4). Cellular appearance varied from fibroblastic-like cells with elongated nuclei and scanty cytoplasm to epithelioid-like oval cells with abundant eosinophilic cytoplasm and round to oval, centrally located nuclei.

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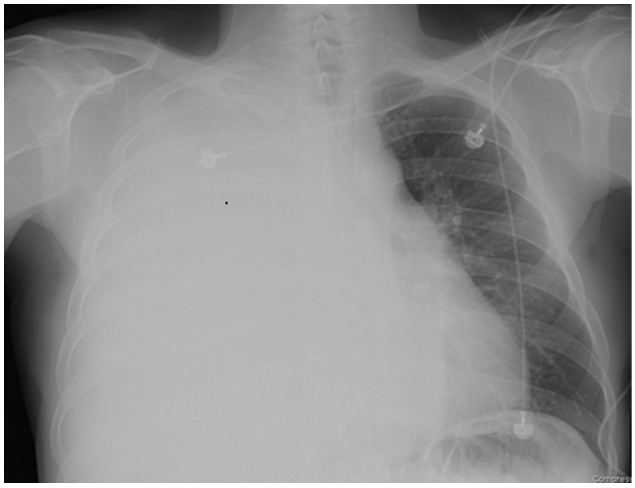

Figure 1 Chest radiograph posteroanterior view shows complete right hemithorax opacification with contralateral mediastinal shift.

Mitoses, cytological atypia, necrosis or haemorrhage were not identified (figure 4).

Immunohistochemical staining was diffusely positive for CD34 (figure 5) and BCL2 (figure 4A; figure 5); it was focally positive for cytokeratin AEI/AE3, and negative for S100-protein and α-smooth muscle actin. On the basis of these morphological and immunohistochemical findings, the diagnosis of SFTP was made.

The patient underwent preoperative angiography and embolisation of the tumour followed by complete resection via an extended posterolateral thoracotomy.

INVESTIGATIONS

- Chest radiograph
- CT chest, abdomen, pelvis
- Percutaneous CT-guided biopsy

DIFFERENTIAL DIAGNOSIS

- Pleural lipoma
- Neurofibroma
- Bronchogenic carcinoma
- Sarcoma

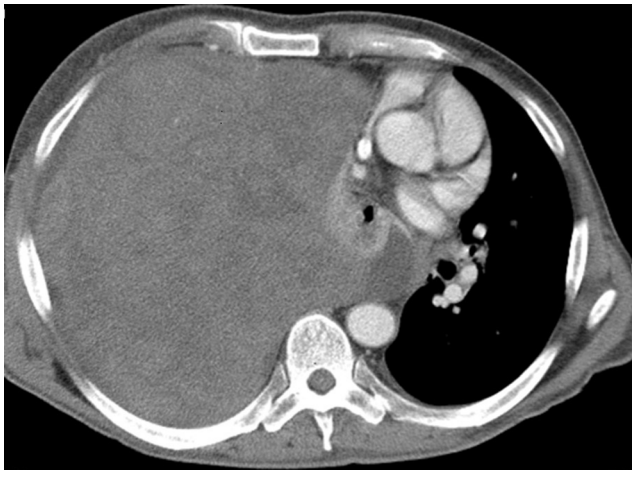

Figure 2 Contrast-enhanced CT of the chest shows a 21×21 cm heterogeneous mass abutting the chest wall with compression and contralateral displacement of mediastinal structures.

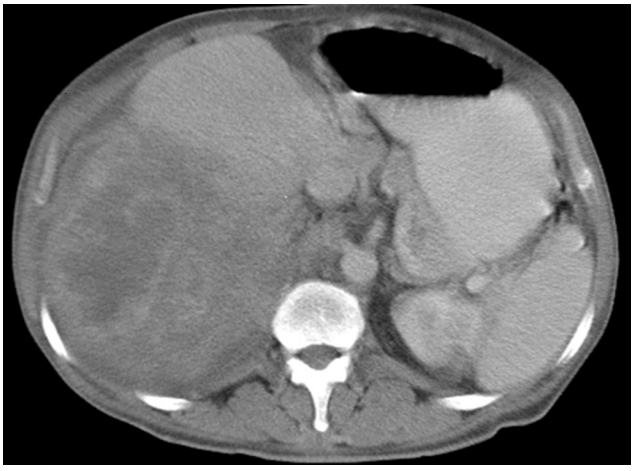

Figure 3 Contrast-enhanced CT of the abdomen/pelvis shows notable inferior and lateral displacement of the liver by a large heterogeneous mass.

- Liposarcoma
- Mesothelioma
- Thymoma

TREATMENT

- Preoperative angiography with embolisation of the tumour
- Tumour resection via thoracotomy

OUTCOME AND FOLLOW-UP

The patient was seen in clinic 4 weeks after undergoing surgical resection. His dyspnoea has markedly improved and his lower extremity swelling had resolved. Follow-up chest radiograph showed near complete re-expansion of the right lung.

DISCUSSION

First described by Klemperer and colleagues in 1931, SFTs are rare neoplasms originally thought to arise from submesothelial connective tissue.[2] A recent review noted approximately 800 cases total reported in the medical literature.[3] Recent immunohistochemical studies have proven SFTs to actually be of mesenchymal origin. A variety of organ systems may be affected, including orbit, lower respiratory tract, peritoneal cavity, heart, salivary glands, liver, breast, soft tissues, retroperitoneum, thyroid and meninges.[4–6] Research does not point to a genetic

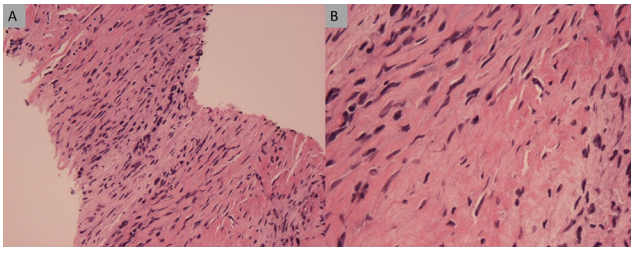

Figure 4 (A) H&E stained tissue shows tumour composed of haphazardly arranged bland-looking spindle cells admixed with thin collagen fibres. (B) Cell appearance varies from fibroblastic-like cells with elongated nuclei and scanty cytoplasm to epitheliod-like oval cells with abundant eosinophilic cytoplasm and round to oval, centrally located, nuclei.

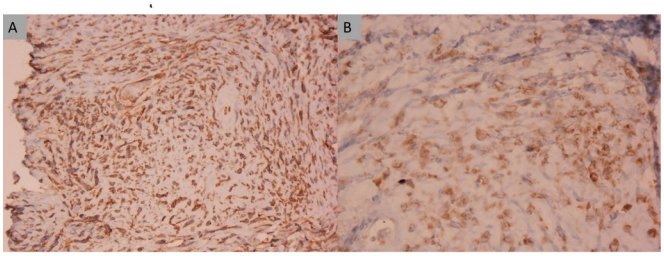

Figure 5 Immunohistochemical analysis demonstrating positive CD34 (A) and BCL2 (B) stains.

predisposition for SFTPs nor does it indicate any relationship to exposure to asbestos, tobacco or other environmental agents.[3]

SFTPs tend to affect adults during the sixth and seventh decades of life and typically present as incidental asymptomatic masses, often quite large in as many as 50% of cases, with concomitant ipsilateral pleural effusion in 17% of cases.[3 7] Larger tumours are generally more likely to present with symptoms while smaller tumours are more commonly asymptomatic.[3] Respiratory symptoms may occur in one-third of patients. These include cough, dyspnoea, pleuritic chest pain, and haemoptysis due to compression of a bronchus.[7–9] Extrathoracic symptoms include hypoglycaemia in 5% of patients and hypertrophic osteoarthropathy in up to 20% of patients, thought to be related to increased production of hyaluronic acid by tumour cells.[8 10 11] In patients with hypertrophic pulmonary osteoarthropathy, which is frequently associated with larger tumours, bilateral arthritis-like symptoms such as oedema of the ankles and swelling of the joints are often displayed.[9] Additional manifestations include weight loss, galactorrhoea and arthritic pain.[10 11] Our patient presented with respiratory symptoms and weight loss as well as lower extremity oedema.

SFTPs are often incidentally found on plain chest radiography or CT. However, the appearance on plain chest radiography is non-specific and should prompt further investigation.[3] Contrast-enhanced chest CT is considered the key initial diagnostic modality that can accurately visualise the extent of tumour, its location and morphology, and assist with preoperative surgical evaluation.[9] Although there are no typical radiological features, small SFTPs are usually well defined, lobular in shape and homogenous in density, while larger tumours are most commonly seen as diffuse pleural opacities which are heterogeneous in density.[9] Malignant and benign SFTPs cannot be distinguished by imaging studies alone; however, increased heterogeneity, necrosis, cystic changes, size greater than 10 cm and pleural effusion are indicative of possible malignant transformation.[12]

Additional radiographic modalities which can occasionally assist in the evaluation of SFTPs include MRI and positron emission topography (PET). Although MRI is of limited utility for the assessment of most pleural tumours, it can provide valuable information relating to the relationship of the tumour to adjacent mediastinal or major vascular structures, or the presence and extent of vertebral/foraminal involvement for large posterior tumours.[3] PET scanning has been utilised in select cases in an effort to help distinguish between benign and malignant tumours; however, minimal fluorodeoxyglucose uptake in these cases suggest that PET adds little to the evaluation of SFTPs.[3]

The diagnosis of SFTPs can only be confirmed through histopathological analysis.[3] Histologically, a SFTP is characterised by fibroblast-like cells and connective tissue in varying proportions, often arranged in a 'patternless pattern' as well as a haemangiopericytoma-like pattern.[13 14] Hiraoka and colleagues reviewed the immunochemical findings of 12 patients with SFTP. Specific antibodies studied included CD34, vimentin, keratin, desmin, α-SMA, S-100, Bcl-2, p53, Ki67, and CD117. All specimens were positive for vimentin and a majority were positive for CD34, as has been noted in previous studies.[15] No specimens in this study exhibited reactivity for keratin or S-100, findings also consistent with previous studies.[6] Bcl-2 expression strongly correlated with tumour diameter. Our patient's specimen was CD34 and BCL2 positive, actin and S-100 negative and keratin and AE1/AE3 were focally positive. These immunohistochemical results are vital in distinguishing the SFTP from other spindle cell tumours of the pleura.[3]

SFTPs are now known to show variable malignant potential, which is defined histologically. While standardised criteria have yet to be established, review of several studies shows several common features which generally define malignancy: high mitotic activity, high cellularity with crowding and overlapping of nuclei, pleomorphism and possibly necrosis.[16 17]

Due to their non-specific clinical presentation, SFTPs are often diagnosed in the latter phases of their development when they cause compression, displacement or, less commonly, invasion of the surrounding structures. Complete en bloc surgical resection via thoracotomy is the mainstay of therapy for most localised SFTPs. Resections of giant-sized tumours, such as the one described herein, is more challenging due to poor exposure and visualisation, as well as significant blood supply to the tumour. Preoperative angiography and embolisation of major vessels can make the operation safer by significantly reducing the risk of intraoperative bleeding.[18]

The vast majority of patients undergoing surgical resection of SFTPs have a benign clinical course, with a 10-year disease-free survival of 90%.[6 19] Long-term follow-up after complete resection is advised due the risk of late recurrence of SFTPs.[20] Malignant SFTPs may be curable provided complete resection is achievable. However, local recurrence is possible, with one study reporting a 63% recurrence rate, even with complete resection.[3] There is no data supporting the use of chemotherapy or radiation therapy for patients at high risk for recurrence. Therefore, close follow-up of high-risk patients using chest CT is recommended.[3 17 20]

Learning points

- Larger solitary fibrous tumours of the pleural (SFTPs) can present with pulmonary symptoms, such as cough, shortness of breath and chest pain, although the majority of cases are found incidentally during chest imaging. Characteristic extrapulmonary symptoms include hypoglycaemia and hypertrophic osteoarthropathy.
- Diagnosis of SFTP requires histological examination of an adequate tissue sample and is based on identification of typical morphological features in conjunction with characteristic immunohistochemical findings.
- Surgical resection is the mainstay of therapy for all localised SFTPs. Resection of giant-sized SFTPs can be more challenging, and can potentially be made safer with preoperative angiography and embolisation.

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