CASE REPORT

Solitary fibrous tumour of the pleura masquerading as catecholamine-secreting paraganglioma

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SUMMARY

A 33-year-old African-American woman presented with left-sided chest pain for 2 months before admission. Physical examination revealed no breath sound in the left chest and CT scan of the chest showed total obliteration of the left pleural cavity. The patient also had hypertension and elevated urinary metanephrines, leading to a tentative diagnosis of a catecholamine-secreting paraganglioma. MRI revealed a large, heterogeneous soft tissue mass that occupied the entire left chest cavity, causing displacement of the heart and mediastinal structures to the right. Through a left thoracotomy incision, a tumour weighing 2790 g was removed along with a small portion of adherent lung. The tumour was positive for CD34 but negative for S-100, keratin, desmin and progesterone-receptor, which is consistent with pathological diagnosis of a solitary fibrous tumour of the pleura. The patient remains symptom free 4 years after the operation.

BACKGROUND

Solitary fibrous tumours of the pleura (SFTP) are rare primary tumours of the pleura.¹ They arise from mesenchymal tissue underlying the mesothelial layer of pleura and are benign approximately 80% of the time.² Here, for the first time, we report a case of a gigantic SFTP that, on initial endocrine studies, appeared as a catecholamine-secreting paraganglioma.

CASE PRESENTATION

A 33-year-old African-American woman presented with left-sided chest pain of 2 months duration. Physical examination revealed no breath sounds in the left chest. CT scan of the chest showed total obliteration of the left pleural cavity (figure 1). On further investigations, the patient had elevated urinary metanephrines; however, the radioactive-labelled metaiodobenzylguanidine (MIBG) of total body scintigraphy failed to detect a pheochromocytoma. A tentative diagnosis of catecholamine-secreting paraganglioma in the left pleural cavity was made. She was started on phenoxybenzamine and atenolol in preparation for surgery.

INVESTIGATIONS

MRI revealed a large (maximum dimensions 15.3 cm anteroposterior, 13.6 transverse, 19.5 cm craniocaudal), heterogeneous soft tissue mass occupying the entire left chest cavity, displacing the heart and mediastinal structures to the right.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of solitary pleural fibroma includes solitary pleural metastasis, lipoma, pleural fibrosarcoma, intercostal nerve neurilemoma, organised inflammation and peripheral bronchial carcinoma. In the case described here, catecholamine-secreting paraganglioma was considered in the differential diagnosis of the elevated urinary metanephrine levels and a negative MIBG scan.

TREATMENT

Through a left thoracotomy incision, at the fifth intercostal space, a tumour weighing 2790 g was removed along with a small portion of adherent lung. Sectioning of the tumour revealed firm, pink and white, uniform tissue with areas of whorled appearance (figure 2). On immunohistochemical staining, the tumour was positive for CD34, but negative for S-100, keratin, desmin and progesterone receptor (figure 3). These findings are consistent with pathological diagnosis of an SFTP.

OUTCOME AND FOLLOW-UP

The postoperative recovery period was uneventful and the patient was discharged home with no complications. A CT scan of the chest performed 2 weeks postoperatively, showed re-expansion of the left lung with minimal air space and fibrotic changes. She is symptom free 4 years after the operation with urinary catecholamine in a normal range.

DISCUSSION

Primary tumours of the pleura are divided into two categories: diffuse and solitary. Diffuse tumours, known as diffuse pleural mesotheliomas, are the most common type. They arise from the mesothelium, are associated with asbestos exposure and are highly malignant.³ Solitary tumours or SFTPs are usually benign, without association with smoking or asbestos exposure.⁴ Immunohistochemical studies have shown that solitary tumours likely arise from mesenchymal tissue as they express vimentin, but not cytoplasmic keratins, as found in mesotheliomas. Solitary fibrous tumours may occur in a number of tissues, including the nasopharynx, epiglottis, thyroid, mediastinum, peritoneum and liver, supporting a mesenchymal origin.⁵ SFTPs make up approximately 2.8/100 000 tumours, and less than 5% of pleural neoplasms. Peak presentation occurs in the sixth and seventh decades, although they may affect any age group.⁶

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Figure 1 CT scan of the chest. (A) Before surgery, the tumour completely obliterated the left pleural cavity, displacing the heart and mediastinal structures to the right. (B) After surgery, there is re-expansion of the lung and restoration of the positions of the heart and mediastinal structures.

Clinically, the tumours are often asymptomatic until they become large enough to produce localised symptoms such as chest pain, cough or dyspnoea. Systemic symptoms such as weight loss, sweating, chills and paraneoplastic symptoms such as clubbing of the fingers and hypoglycaemia may also be present.⁴

The majority of SFTP arise from the visceral pleura. Radiological findings often show a sharply delineated, homogeneous, sometimes lobulated, round mass, although opacification of the entire hemithorax may occur in very large tumours. Contrast enhancement is usually intense due to rich tumour vasculature, although areas of non-enhancement due to necrosis, myxoid degeneration or haemorrhage may be present.⁷

The differential diagnoses for SFTP include solitary pleural metastasis, lipoma, pleural fibrosarcoma, intercostal nerve neuroma, organised inflammation and peripheral bronchial carcinoma.⁸ Catecholamine-producing SFTPs have not been reported previously; therefore, in the case described here, catecholamine-secreting paraganglioma was considered in the differential due to the elevated urinary metanephrine levels.

While surgery remains the gold standard for definitive diagnosis of SFTPs, preoperative diagnosis can be made by histological and immunohistochemical analysis through transthoracic cutting-needle biopsy.⁶ Fine-needle transthoracic aspiration

cytology is usually inconclusive because of the presence of acellular and hypercellular portions in the tumour.⁵ Alternatively, undiagnosed thoracic tumours may first be evaluated by video-assisted thoracoscopy.¹ While smaller tumours (<10 cm) have been successfully removed using video-assisted thoracoscopic surgery with fewer postoperative complications and shorter hospital stays, thoracotomy is the preferred method of resection for larger tumours.^{5–7} Resection generally only requires a small margin of normal tissue, and recurrence is rare with complete resection.

Microscopically, SFTPs display uniform, elongated spindle cells with variable amounts of collagen and reticular fibers.^{2–7} On immunohistochemical analysis, SFTPs are negative for keratin, desmin and S-100, and positive for CD34 and vimentin.^{3–4,7}

Criteria for malignant potential of SFTPs include abundant cellularity, greater than 4 mitoses per 10 high-power field, cytological atypia, large necrotic or haemorrhagic areas, pleural effusion, atypical location and invasion of adjacent structures.

Long-term follow-up is advised due to the risk of recurrence and malignant transformation years after surgical resection. Contrast-enhanced CT scan should be performed every 6 months for the first 2 years, then yearly afterwards.⁷

Figure 2 Solitary fibrous tumour: the tumour is composed of spindle cells with bland oval nuclei that are separated from one another by eosinophilic collagen. Vascular spaces are present in the tumour. No necrosis or mitotic figures are present (H&E $\times 200$).

Figure 3 Immunohistochemical staining of solitary fibrous tumour. The tumour is positive for CD34 (brown colour; $\times 200$), and negative for S-100, keratin, desmin and progesterone receptor (not shown).

Learning points

- ▶ Solitary fibrous tumour of pleura can be gigantic, and obliterate the entire pleural cavity.
- ▶ Solitary fibrous tumour of pleura can produce catecholamines.
- ▶ Surgical resection is the treatment of choice.
- ▶ Long-term follow-up after surgical resection is indicated due to the risk of recurrence.

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