Disseminated solitary fibrous tumour of the lung and pleura

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SUMMARY

Solitary fibrous tumours (SFTs) are a heterogeneous group of rare spindle-cell tumours. Classically they presented as a solitary pleural-based mass. Pulmonary parenchymal SFT is rare and multiple bilateral lesions are extremely rare. We present the clinical, imaging and histological features of SFT which are presented as multiple nodular lesions of the lung and pleura with probable distant metastasis.

BACKGROUND

Solitary fibrous tumours (SFTs) were mostly termed hemangiopericytomas in the past. In 1931, Klemperer and Rabin¹ described SFT as a unique entity. SFTs are a rare neoplasm, with fewer than 800 cases reported in the literature.² The majority of them are pedunculated masses with benign histological features.³ Approximately 12% of SFTs are malignant and eventually lead to death through local recurrence or distant metastasis.⁴ According to the WHO, these tumours are mesenchymal neoplasms of subendothelial origin that can be found mostly in the pleura, but also in extraserosal sites such as lung, mediastinum, liver, head and neck and deep soft tissues of the extremities.⁵ These tumours are observed in middle-aged adults between 20 and 70 years (median 50 years) with no gender predilection. They occur occasionally in children and adolescents. Most SFTs behave as slowly growing, painless masses. Large tumours may give rise to compression symptoms or may be the source of paraneoplastic syndrome due to the production of insulin-like growth factor. Usually, they are found incidentally by chest radiography. The behaviour of SFTs is unpredictable. Approximately 10–15% behave aggressively, so long-term follow-up is mandatory.⁶ We reported an extremely rare case of SFT with bilateral lungs and pleural involvement with probable distant metastasis.

CASE PRESENTATION

A 48-year-old woman was admitted with chest pain on the right side and shortness of breath since 3 months. She had no history of smoking or tobacco chewing. She was malnourished with a body mass index of 18 kg/m². Her oxygen saturation in room air was 97%. On examination, there was decreased air entry on the right hemithorax with stony dull note on percussion. The patient had not been previously exposed to asbestos and there was no family history of malignancy or tuberculosis.

Her chest X-ray (figure 1) showed bilateral nodular lesions in the lung parenchyma with pleural effusion on the right. The CT scan revealed

well-defined enhancing nodules with central hypodensity, of varying sizes involving both the lungs. There were also discrete nodular lesions along the mediastinal, fissural and diaphragmatic pleura in addition to loculated pleural effusion on the right (figure 2). Also, there was a well-defined enhancing lesion with central hypodensity and calcification noted in the left gluteus maximus muscle.

The CT-guided biopsy of the right lung nodule was performed and it was reported as SFT. Microscopy of the specimens (figure 3) revealed a tumour composed of spindle cells arranged in storiform pattern, plump ovoid to elongated nuclei with fine chromatin, inconspicuous nucleoli and moderate to abundant pale to eosinophilic cytoplasm. Thin-walled vascular channels are interspersed. There is no increased mitotic activity or necrosis. Immunohistochemistry revealed that the tumour cells are positive for B-cell lymphoma 2 (BCL) and CD34 stains the interspersed blood vessels.

Since SFT does not usually present as multiple nodules a repeat biopsy of another nodule was carried out which again showed similar histology. Flexible bronchoscopy excluded an endobronchial lesion and a transbronchial lung biopsy from the right lung showed evidence of SFT. A pleural fluid analysis revealed an exudative pleural effusion on the right and an unguided pleural biopsy was inconclusive. Medical thoracoscopy with a semi-rigid thoracoscope was carried out which revealed multiple nodular lesions on the parietal pleura (figure 4). Biopsy of these lesions revealed histology picture similar to SFT. Since three out of the four different biopsies revealed similar histology, a diagnosis of multiple metastatic SFT was made. A positron emission—CT examination revealed uptake in the nodular lesions of lungs and pleura with a single hot lesion in the left gluteus maximus muscle with uptake similar to the lesion in lung. We did not do biopsy from the gluteus maximus muscle because there was already three sites biopsy proven for SFT.

DISCUSSION

Pulmonary SFTs resemble pleural SFTs. There are two theories regarding their origin. The first one proposes that the neoplasm arises from the subpleural mesenchyma which is in continuity with interlobular septa, and the second proposes that they arise directly from the submesothelial elements of the lung parenchyma.³ SFTs can develop from the visceral pleura as well as the parietal pleura.

It is well-known that SFT of the lung and pleura are incidentally discovered on chest X-ray

To cite: Singh RK, Thangakunam B, Isaac BTJ, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009362

Figure 1 Chest X-ray bilateral multiple nodules with the right pleural effusion.

examination because these neoplasms often have a silent clinical course for several years.^{2 3} It has been described in all ages, but the peak incidence is in the sixth and seventh decades of life.¹ The larger the tumour, the more likelihood of causing symptoms.^{3 5} Systemic symptoms such as weight loss, nocturnal sweating, chills, weakness, digital clubbing, hypertrophic osteo arthropathy (HOA), and hypoglycaemia have also been reported and goes by the name Pierre Marie-Bamberg syndrome is HOA and not the entire constellation of symptoms and HOA is probably related to abnormal hyaluronic acid production.^{2 4} It is related to the abnormal production of hyaluronic acid by the tumour cells and affects up to 20% of patients. In less than 5% of cases, SFT can secrete insulin-like growth factor 1 which causes refractory hypoglycaemia.^{1 4} Unusually SFT may present with leg oedema and dyspnoea caused by a large tumour compressing the right atrium and the inferior vena cava as reported by Shaker *et al.*⁷ Our index patient presented with cough and shortness of breath.

Definitive diagnosis of the lesion is based on histological examination of the tumour and on their immunohistochemical characteristics (keratin, negative; S-100 negative; vimentin, positive). All previous reports have agreed that the efficacy of therapeutic strategy depends on the completeness of the tumour resection.⁴ Adjuvant therapy appeared to play a role in recurrent or systemic disease, but its benefit is undefined. SFTs have an unpredictable course, depending on their potential to turn malignant. They often recur on the site of the resection or

Figure 2 CT of the thorax bilateral multiple nodular lesion with pleural effusion on the right.

Figure 3 Microscopy of the specimens revealed a tumour composed of spindle cells.

appear as metastasis. Therefore, it is mandatory to retain an annual follow-up with regard to all compartments.

INVESTIGATIONS

Immunohistochemistry was carried out which revealed that the tumour cells were positive for BCL-2. The CD34 stains the interspersed blood vessel. The interstitial cells of Cajal (ICC) markers showed that the tumour cells are locally positive for CD99 (figure 5) and BCL-2.

CONCLUSION

Generally, SFT is a localised benign tumour. It is curable by a careful and complete resection. But it is important to remember that rarely it may have an malignant clinical course with extensive metastasis as in our case. In our case it was not possible to resect the lesions, as they are widespread involving the lungs as well as the pleura.

Figure 4 Thoroscopic view of the parietal pleura showing multiple nodules.

Figure 5 Interstitial cells of Cajal marker showing tumour cells locally positive for CD99.

Learning points

- ▶ Solitary fibrous tumors though localised and benign, can occasionally present with multiple metastatic lesions.
- ▶ Thoracoscopic pleural biopsy has better yield than closed pleural biopsy in pleural involvement of solitary fibrous tumor.
- ▶ The CD34 stains and the interstitial cells of Cajal (ICC) markers being positive for CD99 and BCL-2 are diagnostic of solitary fibrous tumor.

Acknowledgements The authors are grateful to Dr Saheer, Fellow Pulmonary Medicine Department, Christian Medical College, Vellore, for their continuous encouragement to write this article.

Contributors AG has provided histopathological picture and other authors were involved in editing the article.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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