CASE REPORT

Synchronous sclerosing haemangiomas of the lung: an unusual presentation of a rare tumour

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SUMMARY

Pulmonary sclerosing haemangioma is a rare lung tumour with a preponderance for Asian oriental women. Typically a well-circumscribed, solitary lesion with benign characteristics, there are concerns of malignant potential including regional lymph node metastases. Four histological subtypes exist. Diagnosis can be challenging as patients are usually asymptomatic and radiological findings are non-specific. Surgical resection alone is diagnostic and therapeutic. We present a case of this rare tumour with an uncommon presentation as two synchronous tumours initially misdiagnosed as a pulmonary adenocarcinoma.

BACKGROUND

Pulmonary sclerosing haemangioma (SH) is a rare lung tumour first described by Liebow and Hubbell in 1956.¹ It presents predominantly in women in the fourth or fifth decade with a higher reported incidence in Asian orientals. SH typically appears as a well-circumscribed, solitary, benign lung lesion but some reports suggest a malignant potential. Histologically, four main patterns exist: papillary, sclerotic, solid and haemorrhagic. Patients are usually asymptomatic and radiological findings non-specific hence diagnosis can be challenging. Surgical resection alone is curative, but what constitutes the optimal surgical management is yet to be established. We present a rare case of two synchronous tumours in a patient in whom the initial diagnosis was suggestive of a primary lung adenocarcinoma.

CASE PRESENTATION

Case report

A 59-year-old Chinese female non-smoker presented with a 1-week history of worsening cough and fever. A chest x-ray (CXR) showed mild haziness in the right lung. CT scan revealed two small well-circumscribed lesions in the medial segment of the right lung middle lobe (figure 1). The larger mass which measured 1.8×2 cm was metabolically active on positron emission tomography (PET) imaging with a standardised uptake value (SUV) of 2.8. Histology of a CT-guided biopsy was suggestive of a primary adenocarcinoma with positivity for immunohistochemical (IHC) markers thyroid transcription factor-1 (TTF-1) and cytokeratin-7 (CK-7). Preoperative staging was T4N0 M0 (stage IIb) and a middle lobectomy was performed uneventfully.

Postresection histology revealed two separate circumscribed, unencapsulated nodules exhibiting features of SH. There were two cell types present: cuboidal surface cells and round stromal cells. Both

nodules demonstrated predominantly haemorrhagic and papillary histological patterns, with areas of sclerosis and solid growth patterns. IHC studies demonstrated stromal cells expressing TTF-1 but were negative with CK-7. In contrast, the surface cuboidal cells were positive with both TTF-1 and CK-7, supporting an eventual diagnosis of two independent synchronous SH with mixed haemorrhagic, papillary, sclerosing and solid growth patterns (figure 2).

DISCUSSION

Until recently, the histogenesis of pulmonary SH was deemed controversial. Historically it was thought to be vascular in origin owing to prominent angiomatoid features but contemporary IHC studies and molecular findings suggest the tumour may arise from incompletely differentiated respiratory epithelium.^{2 3}

Figure 1 (A) CT thorax scan demonstrating two well circumscribed lesions in the medial segment of the middle lobe. (B) CT thorax scan demonstrating two well circumscribed lesions in the medial segment of the middle lobe.

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Figure 2 Sclerosing haemangioma: (A) papillary pattern; (B) solid and sclerotic pattern (arrowhead); (C) simple and complex papillae lined by cuboidal surface cells with hyaline collagen within the stalks, presence of foam cells noted; (D) round stromal cells and surface cuboidal cells, some showing nuclear clearing and intranuclear inclusions; (E) surface cells express , cytokeratin 7, which is negative in stromal cells; (F) surface and stromal cells, both express , thyroid transcription factor 1.

SH is most common in women, with a female : male ratio of 5:1. The female predilection is attributed to progesterone receptors in the nuclei of pulmonary SH cells.⁴ SH is a rare benign lung tumour, most common in Asia and hence should feature high on the list of differential diagnoses of any well-defined opacity on the CXR in any middle-aged Chinese woman.⁵ It is usually an incidental finding as patients are typically asymptomatic although some report chronic cough, haemoptysis and chest pain. Fever and expectoration are less reported. Our patient complained of a chronic cough and fever. Rare cases are reported to have regional lymph node metastases; however, metastases do not appear to affect long-term survival.³

A differential diagnosis of SH should include adenocarcinoma, tuberculoma, carcinoid tumour, hamartoma, malignant teratoma, angiosarcoma, cavernous haemangioma and arteriovenous malformations.⁶ The diagnosis as this case illustrates can prove challenging. Typically, a CXR would reveal a well-defined coin lesion in the lung parenchyma and CT imaging usually reveals a peripheral, solitary mass. Our patient had two proven separate SH tumours which is a rarely reported phenomenon and this may have contributed to the initial misdiagnosis as a malignant adenocarcinoma. Importantly, CT features of an SH are non-specific and hence rarely diagnostic. Fludeoxyglucose (FDG)-PET/CT evaluation of SH is limited by a high false-positive rate in part owing to the increased SUV seen with larger tumours.⁵⁻⁷ In this patient the larger lesion was metabolically active on PET scanning and falsely positive for malignancy. The precise role of PET in the diagnosis of pulmonary SH remains undefined and presently results must be interpreted with caution. Additional use of diffusion-weighted imaging (DWI) MRI may aid PET diagnosis.⁸ Lesions with an SUV >2.5 suggestive of malignancy on PET can be further assessed with DWI which can differentiate malignant lesions owing to greater cellularity.

Preoperative histological diagnosis of SH may also prove difficult as illustrated with this case as the needle biopsy specimen

showed features suggestive of adenocarcinoma. In one study, 0 of 24 cases were correctly diagnosed as SH preoperatively after histological analysis.⁹ Surgery is curative and it is the only effective treatment for pulmonary SH. Segmentectomy or wedge resection should be first considered for cases without a definite preoperative diagnosis.⁹ Intraoperative frozen section histological analysis can help exclude malignancy thereby limiting the extent of resection. However, in cases where even an intraoperative frozen section analysis is equivocal, anatomic resection and systematic lymph node dissection is recommended.

Pulmonary SH poses a significant diagnostic challenge as this case illustrates. PET alongside DWI may reduce misdiagnosis as a malignancy. Preoperative biopsy can be misleading and an intraoperative frozen section may be required. Parenchymal sparing lung resection, the treatment of choice is both diagnostic and therapeutic. The case described demonstrates that an atypical presentation with multiple lesions should not exclude the possibility of pulmonary SH.

Learning points

- ▶ Pulmonary sclerosing haemangioma (SH) is a rare benign lung tumour that is usually a solitary lesion, however, multiple or synchronous lesions should not exclude the diagnosis.
- ▶ Diagnosis can be challenging owing to non-specific clinical and radiological features and the lesion can mimic malignancy.
- ▶ Pulmonary SH should be strongly considered in any Asian oriental female non-smoker with a solitary pulmonary nodule.
- ▶ Parenchymal sparing lung resection, the treatment of choice is both diagnostic and therapeutic.

Competing interests None.

Patient consent Obtained.

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