

CASE REPORT

Endobronchial benign nerve sheath tumour presenting with significant shortness of breath and haemoptysis

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Abstract

Peripheral nerve sheath tumours are rare within the thoracic cavity, with non-specific presentation. A 29-year-old patient presented with shortness of breath, cough, haemoptysis and recurrent chest infections. Suspicion of a primary lung carcinoma or a neuroendocrine tumour was raised following a CT and PET-CT. An endobronchial tumour suggested on histology a diagnosis of benign nerve sheath tumour, with positive staining for S100, CD56 and CD34. Following lung resection, the patient complained of fatigue and developed subcutaneous erythematous nodules on the anterior right chest, which raised the suspicion for a differential diagnosis of neurofibromatosis type I. The nodules resolved spontaneously within two weeks and the diagnosis of neurofibromatosis was ruled out on subsequent magnetic resonance imaging head and chest.

BACKGROUND

Peripheral nerve sheath tumours (PNST) are relatively rare within the thoracic cavity, and when present, are most commonly found within the posterior mediastinum [1]. Benign PNSTs such as neurofibromas, perineuriomas and ganglioneuromas can be endobronchial, while schwannomas are usually located in the posterior mediastinum [2].

The clinical presentation is non-specific, but it can be more characteristic in cases of neurofibromatosis type I and type II [1, 3]. Malignant intrathoracic PNSTs are rare, become symptomatic following compression of mediastinal structures and show an aggressive behaviour with poor prognosis [2].

Soft tissue tumours are generally assessed radiologically by magnetic resonance imaging, however, in cases of type I neurofibromatosis, positron emission tomography (PET) has been used to distinguish between benign and malignant lesions prior to planning appropriate treatment [3].

Histologically, schwannomas can be identified by their strong staining for S100 protein, whilst neurofibromas generally react less intensely for this antibody and contain a subpopulation of CD34 positive fibroblasts [2, 4].

CASE REPORT

A 29-year-old male university lecturer presented to the respiratory outpatient clinic with shortness of breath, cough, haemoptysis and recurrent chest infections. The clinical examination was unremarkable. The patient had a history of asthma, with unlimited exercise tolerance and no history of smoking or asbestos exposure. An incidental finding of a right hilar mass was detected on plain chest x-ray. Following the initial assessment, the patient underwent a CT and PET CT, which showed a well circumscribed intraparenchymal mass with an endobronchial component, which had low fluorodeoxyglucose (FDG) uptake (Fig. 1).

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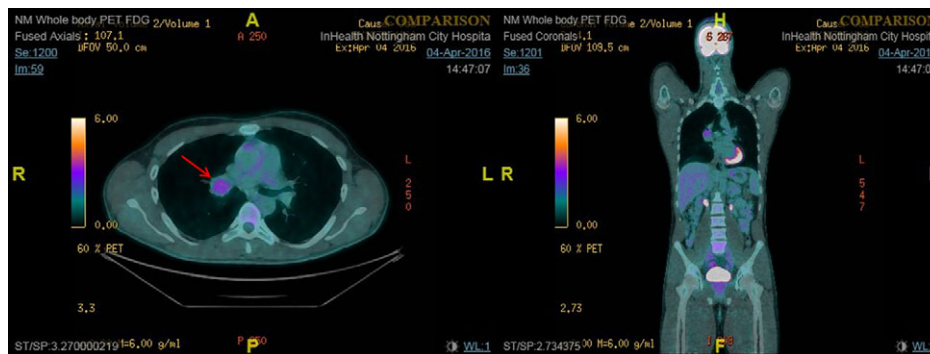


Figure 1: Fused axial and coronal sections from PET CT showing low FDG uptake ($SUV_{max}=3.7$) in the $28 \times 29 \text{ mm}^2$ right hilar mass, without any abnormal distant uptake.

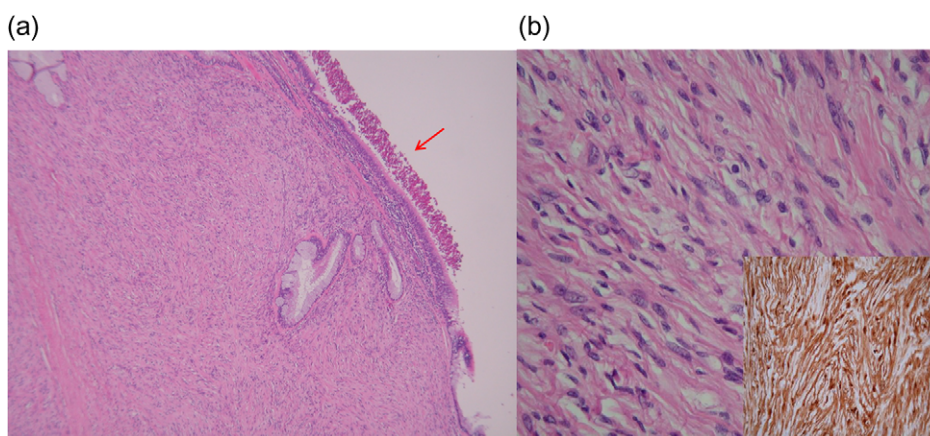


Figure 2: Low power view showing bland spindle cells encroaching upon the bronchial wall epithelium (indicated by red arrow) (a). High power view of delicate spindle cells in a lightly collagenous stroma. The inset image shows the immunoperoxidase positive staining for S100 protein (b).

The patient subsequently underwent rigid and flexible bronchoscopies, showing an endobronchial tumour with satisfactory margins for a sleeve lobectomy. Biopsies from the endobronchial mass suggested a diagnosis of benign nerve sheath tumour. The patient underwent a video-assisted thoracoscopic surgery exploration, followed by open sleeve right upper lobectomy, with the tumour positively identified in the resected lobe.

Histology showed a spindle cell lesion comprised of cells arranged in a haphazard fashion with small amounts of eosinophilic cytoplasm and bland monomorphic nuclei. The tumour was strongly positive on immunohistochemistry for S100 protein and also showed positivity for CD56 and glial fibrillary acidic protein (GFAP), with a subpopulation of CD34 positive cells. The morphological and immunohistochemical findings were unusual and reminiscent of so-called gastrointestinal schwannoma, but lacking the characteristic lymphoid aggregates and inflammatory component. The features were, nevertheless, best characterized as a benign schwannoma and the strongly positive immuno-staining for S100 protein supported this diagnosis (Fig. 2).

Post-operatively the patient complained of fatigue and developed subcutaneous erythematous nodules on the anterior right chest, which raised the suspicion of neurofibromatosis type I. The nodules resolved spontaneously within 2 weeks, with no new changes on the follow-up chest CT scan. The diagnosis of neurofibromatosis type I was ruled out following

genetic testing. No recurrence was identified at one year and the patient was discharged from follow-up.

DISCUSSION

The incidence of pleuropulmonary benign nerve sheath tumours is very low outside the posterior mediastinum [2], which makes diagnosis difficult but no less important in ensuring the optimal management and treatment of these patients. Intrathoracic PNSTs are usually benign. However, malignant forms, if present, are more likely to be associated with neurofibromatosis type 1 [1]. The atypical endobronchial location of the benign nerve sheath tumours could correspond to the bronchial vagus nerve branches [3]. In this case, full excision was essential to confirm the diagnosis and resolve the presenting symptoms. The radiological features and the histological characteristics, when taken together, help in accurately identifying these tumours [2].

As in this case, patients tend to be asymptomatic initially due to the slow growing nature of the tumours [4]. Symptoms result from a compression effect of the enlarging tumours, although hemoptysis is extremely rare [2]. Reports of other cases of mediastinal and pleuropulmonary PNSTs, suggest that complete surgical resection is the optimal management with good prognosis and no risk of recurrence [5].

In conclusion, we have described a case of an endobronchial benign PNST with intraparenchymal extension that presented with unusual but significant symptoms, where histological and

radiological diagnosis coupled with surgical resection ensured optimal treatment and good outcome.

CONFLICT OF INTEREST STATEMENT

None declared.

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