

Endobronchial sclerotic neurofibroma: a very rare benign tumour

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Introduction

A 69-year-old man, with a 66 pack-year smoking history and a background of alcohol-induced liver cirrhosis, peripheral vascular disease and dilated cardiomyopathy, presented with severe normocytic anaemia. Computed tomography of the chest, abdomen and pelvis revealed an indeterminate nodule in the bronchus intermedius. Fiberoptic bronchoscopy showed a polypoid lesion with vascular appearance; this was combined with rigid bronchoscopy under general anaesthetic to remove the lesion with rigid forceps. Histology identified a sclerotic neurofibroma staining positively for S100 and LP10. The patient is due for follow up in 6 months with a computed tomography scan of the chest to ensure that there is no tumour recurrence.

Discussion

Endobronchial neurofibromas are exceedingly rare benign lesions, most commonly originating at the trachea (Boland et al, 2015). This article presents the first occurrence of the sclerotic variant of endobronchial neurofibroma and the approach used in its definitive investigation and management. Primary pulmonary tumours of neurogenic origin such as peripheral nerve sheath tumours are extremely rare, accounting for less than 0.2% of all lung tumours (Roviaro et al, 1983). Intrathoracic peripheral nerve sheath tumours are usually benign and are commonly found in the posterior mediastinum as schwannomas, with female preponderance (Roviaro et al, 1983). Malignant intrathoracic peripheral nerve sheath tumours may rarely occur and can be associated with neurofibromatosis type 1 (Boland et al, 2015).

Case report

A 69-year-old man, who was an ex-smoker with a 66 pack-year history, was referred to haematology with recurrent episodes of severe normocytic anaemia requiring blood transfusions, presenting symptomatically as shortness of breath and fatigue. Gastrointestinal investigations were unremarkable. Past medical history included alcohol-induced liver cirrhosis with a Child–Pugh score A, peripheral vascular disease and dilated cardiomyopathy. There was no history of industrial exposure or tuberculosis contact.

Computed tomography of the chest, abdomen and pelvis was arranged to rule out sinister pathology. This showed an indeterminate nodule in the bronchus intermedius, and he was referred on the 2-week wait lung cancer pathway for further evaluation (Figure 1). He denied any respiratory symptoms of note; no cough or haemoptysis, breathing was unremarkable, and his exercise tolerance was limited to 300 yards because of intermittent claudication.

Fiberoptic bronchoscopy was arranged and this showed a polypoid lesion with vascular appearance at the secondary carina projecting into bronchus intermedius. The patient was referred to the interventional pulmonology team for combined rigid and fiberoptic bronchoscopy under general anaesthetic; a solitary lesion was noted at the orifice of the bronchus intermedius, which was removed using rigid biopsy forceps (Figure 2). No significant bleeding was noted post biopsy and the patient recovered postoperatively without complications.

Histopathology showed an 8x6x5 mm pedunculated lesion with a 2 mm diameter short stalk and histology of a sclerotic neurofibroma with fibroblasts and collagen. Tissue stained positively for S100 (Figure 3a) and LP10 (Figure 3b); staining was negative for markers CK7, Cam5.2 and CD34.

Six-month follow up with a computed tomography scan of the chest has been planned to ensure there has been no tumour recurrence.

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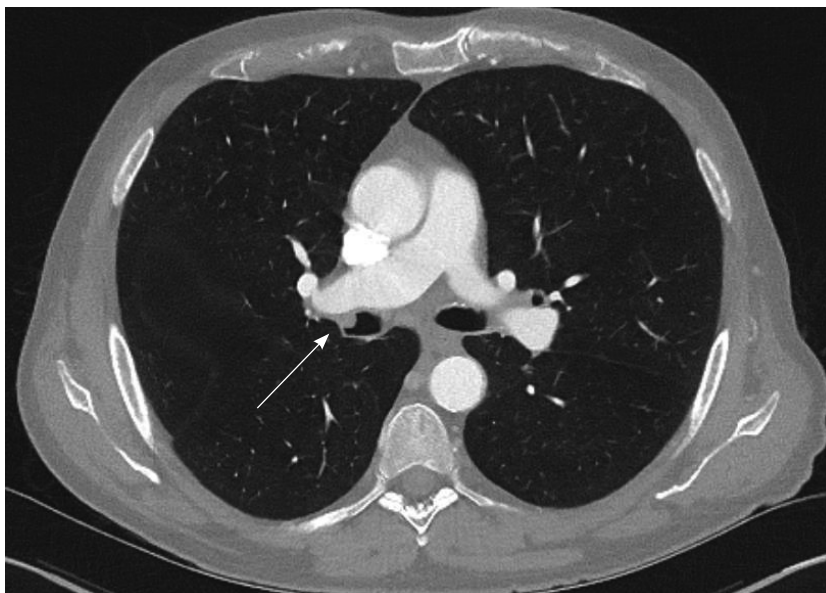


Figure 1. Computed tomography scan of lesion at the bronchus intermedius.



Figure 2. Bronchoscopic image of lesion.

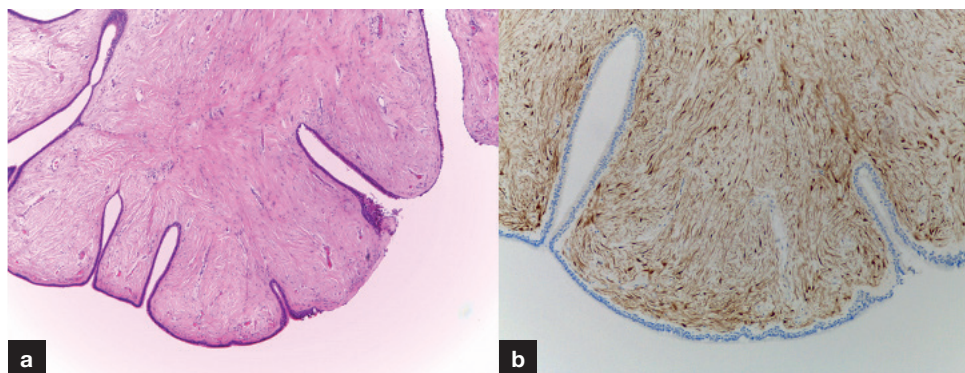


Figure 3. Pedunculated lesion with tissue staining positively for (a) S100 and (b) LP10, showing spindle-shaped cells, scattered fibroblast and collagen.

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Learning points

- Endobronchial sclerotic neurofibromas appear highly vascular and require extra caution during bronchoscopic intervention.
- Experienced interventional pulmonology and thoracic surgical input should always be sought in these cases.
- A combined approach of rigid and fibreoptic bronchoscopy allows better control of the airway and the significant risk of bleeding.

Benign endobronchial peripheral nerve sheath tumours are less common and may relate to the bronchial vagus nerve branches (Okamoto et al, 2013). They include benign schwannomas, neurofibromas, neurilemmomas, perineuroma and ganglioneuroma, with good prognosis following complete resection and no known risk of recurrence (Roviaro et al, 1983; Boland et al, 2015). Most patients are asymptomatic at presentation; depending on the tumour size and degree of airway obstruction, patients may report breathlessness and/or cough. Endobronchial tumour growths resulting in partial or complete obstruction can also cause distal lung collapse and post-obstructive pneumonitis (Rolston and Nesher, 2018). Fibreoptic bronchoscopy can be used for initial evaluation of the lesion. A combined approach with rigid and fibreoptic bronchoscopy, along with other adjuncts such as argon plasma coagulation and cryotherapy, allows better control of the airway and any potential significant bleeding (Feng et al, 2013). Early, prompt involvement of the interventional pulmonology team may enable bronchoscopic resection of benign endobronchial tumours and avoid the need for surgical resection.

To the best of the authors' knowledge, this is the first reported occurrence of the sclerotic variant of an endobronchial neurofibroma. Cutaneous involvement has previously been cited with histological features of predominantly fibroblasts and collagen, rare mast cells and frequent bland S100 positive spindle cells with slender pointed-end nuclei (González-Vela et al, 2006; Nakashima et al, 2008; Rodriguez et al, 2012).

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References

- Boland JM, Colby TV, Folpe AL. Intrathoracic peripheral nerve sheath tumors—a clinicopathological study of 75 cases. *Hum Pathol*. 2015;46(3):419–425. <https://doi.org/10.1016/j.humpath.2014.11.017>
- Feng J, Mao T, Zhao Y et al. Endotracheobronchial neurofibroma treated by right carinal pneumonectomy. *Ann Thorac Surg*. 2013;95(2):e35–7. <https://doi.org/10.1016/j.athoracsur.2012.07.083>
- González-Vela MC, Val-Bernal JF, Gonzalez-Lopez MA et al. Pure sclerotic neurofibroma: a neurofibroma mimicking sclerotic fibroma. *J Cutan Pathol*. 2006;33(1):47–50. <https://doi.org/10.1111/j.0303-6987.2006.00392.x>
- Nakashima K, Yamada N, Yoshida Y, Yamamoto O. Solitary sclerotic neurofibroma of the skin. *Am J Dermatopathol*. 2008;30(3):278–280. <https://doi.org/10.1097/DAD.0b013e3181684525>
- Okamoto J, Kubokura H, Ishii H, Usuda J. Mediastinal neurofibroma originating from the pulmonary branch of the right vagus nerve in a patient without von recklinghausen disease. *Thorac Cardiovasc Surg Reports*. 2013;2(1):029–031. <https://doi.org/10.1055/s-0033-1348477>
- Rodriguez FJ, Folpe AL, Giannini C, Perry A. Pathology of peripheral nerve sheath tumors: diagnostic overview and update on selected diagnostic problems. *Acta Neuropathol*. 2012;123(3):295–319. <https://doi.org/10.1007/s00401-012-0954-z>
- Rolston K, Nesher L. Post-obstructive pneumonia in patients with cancer: a review. *Infect Dis Ther*. 2018;7(1):29–38. <https://doi.org/10.1007/s40121-018-0185-2>
- Roviaro G, Montorsi M, Varoli F, Binda R, Cecchetto A. Primary pulmonary tumours of neurogenic origin. *Thorax*. 1983;38(12):942–945. <https://doi.org/10.1136/thx.38.12.942>