



## Case report

## Intrathoracic ganglioneuroma presenting as an endobronchial mass



Brandon T. Nokes<sup>a</sup>, Coralie P. Baumann<sup>b</sup>, Kristopher W. Cummings<sup>c</sup>, Brandon T. Larsen<sup>d</sup>,  
Rodrigo Cartin-Ceba<sup>b</sup>, Karen L. Swanson<sup>b,\*</sup>

<sup>a</sup> The Department of Internal Medicine, Mayo Clinic, Scottsdale, AZ, United States

<sup>b</sup> The Division of Pulmonary Medicine, Mayo Clinic, Scottsdale, AZ, United States

<sup>c</sup> The Department of Radiology, Mayo Clinic, Scottsdale, AZ, United States

<sup>d</sup> The Department of Laboratory Medicine and Pathology, Mayo Clinic, Scottsdale, AZ, United States

## ARTICLE INFO

## Article history:

Received 6 July 2017

Received in revised form

7 August 2017

Accepted 22 August 2017

## Keywords:

Ganglioneuroma

Endobronchial mass

Intrathoracic

Peripheral nerve sheath tumors

## ABSTRACT

Peripheral nerve sheath tumors (PNST) are exceedingly rare, especially outside of the posterior mediastinum. These tumors represent less than 1% of pulmonary tumors. Very few pulmonary PNSTs are ganglioneuromas. We present a case of a ganglioneuroma presenting as an endobronchial mass.

**Case presentation:** An 80 year old male was seen in pulmonary clinic for routine cancer screening. He had a 60-pack year smoking history. CT evaluation noted a 1cm right lower lobe endobronchial lesion. This lesion was present since 2012 and had slightly increased in size since that time from 8mm (Figure 1). The lesion was further assessed using virtual bronchoscopy (Figure 2). Bronchoscopy revealed an obstructing lesion, which was completely excised with the snare (Figure 3). Pathology revealed well-circumscribed tumor consisting of nests and trabeculae of round/polygonal cells with granular eosinophilic and basophilic cytoplasm. The tumor was chromogranin, synaptophysin, S-100, pancytokeratin, SOX10, and TTF-1 positive, consistent with a ganglioneuroma.

**Discussion:** Aside from a solitary article regarding 75 patient samples (which included only one ganglioneuroma) only a small number of intrathoracic PNSTs have been reported. Only a single case report of an endobronchial ganglioneuroma has been reported. Each of these lesions were benign, and detected on routine imaging evaluations.

**Conclusions:** An intrapulmonary endobronchial location for a PNST is an exceedingly rare presentation of an already uncommon pathology.

© 2017 The Authors. Published by Elsevier Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

## 1. Introduction

Although common in the posterior mediastinum and other somatic soft tissue sites, peripheral nerve sheath tumors (PNST) are exceedingly rare in the lung. These tumors represent far less than 1% of pulmonary tumors, of which ganglioneuromas are even rarer and seldom suspected. We present a case of a ganglioneuroma presenting as an endobronchial mass.

## 2. Case presentation

An 80-year-old male was seen in pulmonary clinic for routine

cancer screening. He had a 60 pack-year smoking history. Computed tomographic evaluation revealed a 1-cm right lower lobe endobronchial lesion. This lesion was present since at least 2012 and slightly increased in size since that time (Fig. 1). The lesion was further assessed using virtual bronchoscopy (Fig. 2). Bronchoscopy revealed an obstructing lesion, which was completely excised with the snare (Fig. 3). Pathology revealed a well-circumscribed submucosal proliferation of Schwannian spindle cells admixed with mature ganglion cells, displaying immunoreactivity for chromogranin, synaptophysin, and S-100 protein in both components, along with SOX-10 expression in the Schwannian component and focal expression of TTF-1 in the ganglion cells, supporting a diagnosis of ganglioneuroma (Fig. 4).

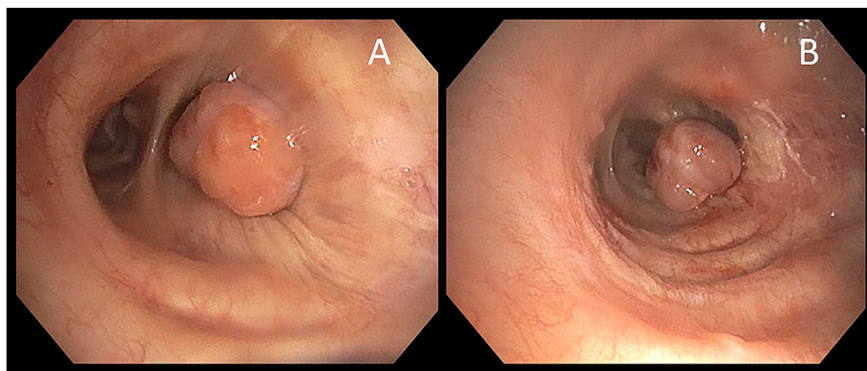
Following removal of the ganglioneuroma, our patient continues to do well 6 months later. He recently presented to our Internal Medicine clinic for routine follow-up, and is in his usual state of health without any new symptomatology following

\* Corresponding author. The Division of Pulmonary Medicine, Mayo Clinic, Scottsdale, AZ, United States.

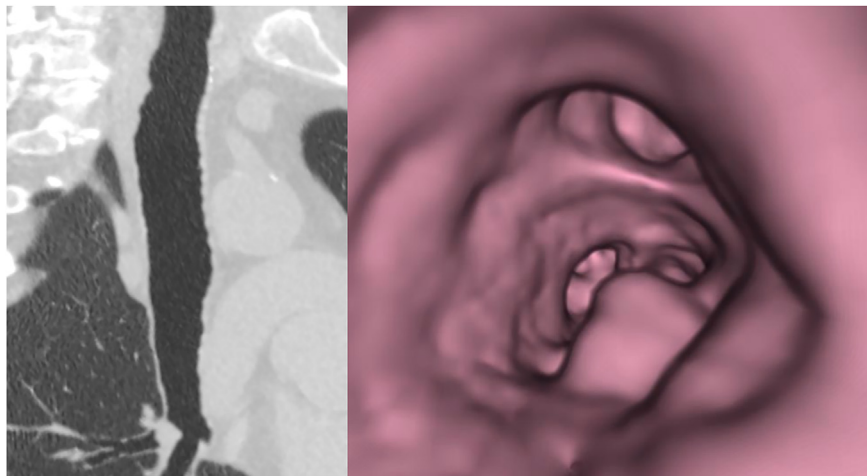
E-mail addresses: [Nokes.Brandon@mayo.edu](mailto:Nokes.Brandon@mayo.edu) (B.T. Nokes), [swanson.karen@mayo.edu](mailto:swanson.karen@mayo.edu) (K.L. Swanson).



**Fig. 1.** Axial CT images at lung window demonstrate only minimal increase in size from 8mm to 10mm of a solitary endobronchial nodule at the origin of the right lower lobe superior segmental bronchus.



**Fig. 2.** Oblique coronal multiplanar reconstruction image (A) demonstrates the position of the circumscribed, endobronchial nodule at the origin of the right lower lobe superior segmental bronchus. Virtual bronchoscopic image from multiple detector CT reconstruction demonstrates the endoluminal lesion from the view of the right lower lobe bronchus.



**Fig. 3.** Large endobronchial lesion at the take off of the superior segment take off. Using a snare the endobronchial abnormality was removed in its entirety. The superior segment was widely patent.

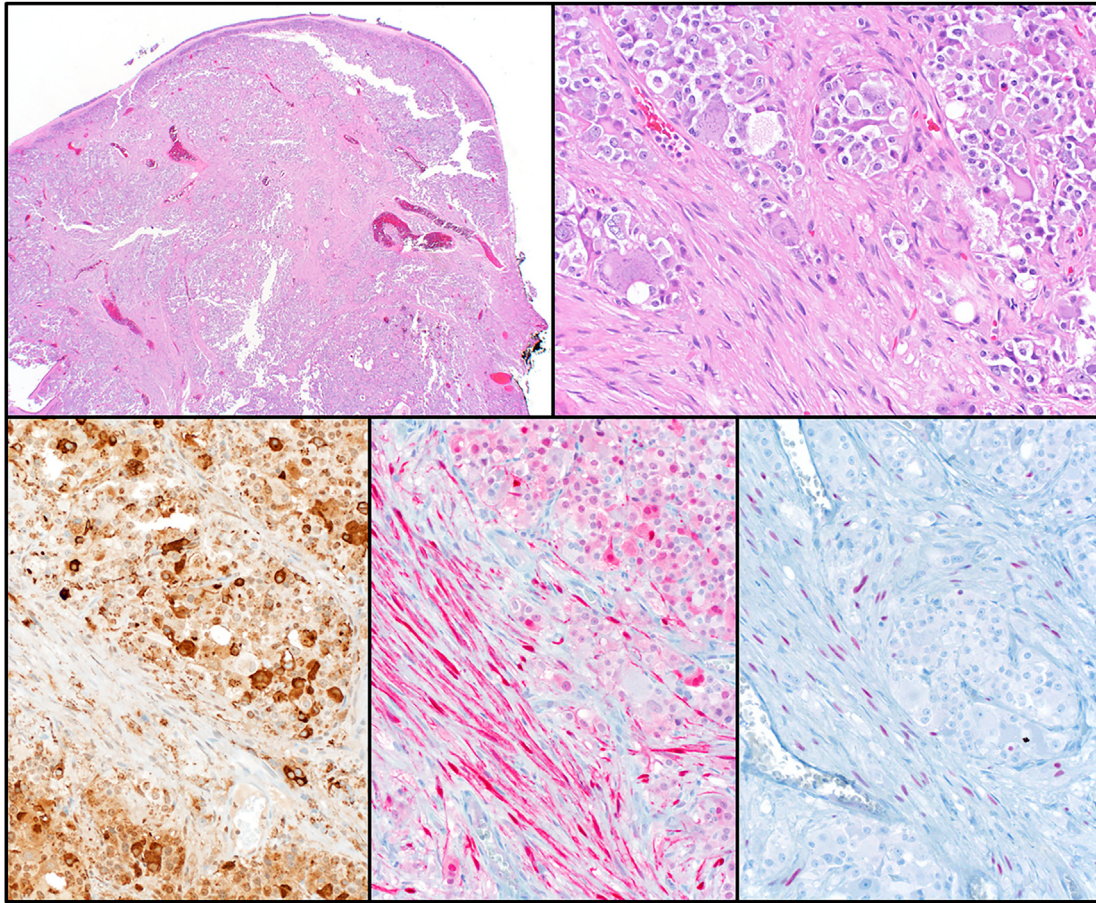
endobronchial removal of his PNST.

### 3. Discussion

Aside from a solitary article regarding 75 patient samples (which included only one ganglioneuroma) only a small number of intrathoracic PNSTs have been reported [1,2]. Only 2 cases have been reported of an endobronchial ganglioneuroma [1,3]. Each of

these lesions were histologically benign, and detected on routine imaging evaluations, although long-term follow-up is probably warranted given their rarity and limited data in the literature. PNSTs are uncommon within the mediastinum. When these tumors do arise, it is typically within the posterior mediastinum. The PNSTs of the posterior mediastinum are thought to arise from spinal nerves, whereas the PNSTs located elsewhere within the mediastinum or pleuro-parenchymal locations have an unknown etiology.





**Fig. 4.** Representative photomicrographs of the resected endobronchial ganglioneuroma. At low power (20 $\times$  magnification, upper left, H&E), a well-circumscribed polypoid submucosal lesion is apparent. At high magnification (200 $\times$ ), the lesion is composed of bundles of Schwannian spindle cells admixed with nests of mature ganglion cells (upper right, H&E) that both display immunoreactivity for chromogranin (lower left, brown pigment indicates positive reaction) as well as S-100 protein (lower mid, red pigment). SOX-10 immunohistochemistry highlights nuclei in the Schwannian component only (lower right, red pigment). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Of the small group of patients with endobronchial PNSTs, they typically present with cough, low-volume hemoptysis, shortness of breath, or post-obstructive pneumonia.

They may also be asymptomatic, as our patient was. Bronchoscopically, the reported endobronchial ganglioneuromas and PNSTs alike have had a rounded appearance within the airway, and our patient's bronchoscopic imaging is in keeping with this finding [1–3]. Radiographically, other more common lesions which may have similar appearances include: salivary gland tumors (mucoepidermoid/adenoid cystic), endobronchial lipoma and endobronchial hamartoma. They are also benign, but may require tissue sampling for definitive diagnosis. There is no available long-term outcome data for the two reported patients with endobronchial ganglioneuromas, though presumably these lesions impart an unremarkable overall clinical course.

#### 4. Conclusions

There are now three reported cases of endobronchial ganglioneuromas. Our case represents the first in which there is clinical follow-up available thereafter. The differential diagnosis for similarly presenting endobronchial masses includes predominantly benign lesions such as salivary gland tumors. Our report provides

data for an overall benign clinical course for patients with endobronchial ganglioneuroma. Albeit exceedingly rare, ganglioneuromas should be included in the differential diagnosis of endobronchial masses.

#### Author contributions

All authors have contributed equally to the concept and design of this manuscript.

#### Funding

The authors have no financial disclosures and there was no research funding provided for this report.

#### References

- [1] J.M. Boland, T.V. Colby, A.L. Folpe, Intrathoracic peripheral nerve sheath tumors—a clinicopathological study of 75 cases, *Hum. Pathol.* 46 (2015) 419–425.
- [2] S. Markaki, C. Edwards, Intrapulmonary ganglioneuroma. A case report, *Arch. Anat. Cytol. Pathol.* 35 (1987) 183–184.
- [3] M.A. den Bakker, A. Marx, K. Mukai, P. Strobel, Mesenchymal tumours of the mediastinum—part II, *Virchows Arch.* 467 (2015) 501–517.