Case report

Endobronchial schwannoma in adult: A case report

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ABSTRACT

Schwannomas are uncommon benign nerve sheath tumors and often arise in the posterior mediastinum and costovertebral angle. However, endobronchial schwannomas are rare. In fact, there are only a few reported cases in the literature. Here, we describe a case of an endobronchial schwannoma causing obstruction of the right upper lobe bronchus.

1. Introduction

Primary neurogenic tumors of the lung are extremely rare. The incidence was approximately 0.2% of all bronchopulmonary tumors and 2.2% of all tracheobronchial ones [1].

Endobronchial schwannomas, also called primary intrabronchial neurilemmas originate from schwann cells. These tumors generally present late. Symptoms are usually nonspecific and some patients are asymptomatic. We reported a case of an endobronchial schwannoma located at the right upper lobe bronchus in an adult.

2. Case report

A 60-year-old man presented with a chief complains of chest pain and dyspnea for about 3 months. His medical history included hypertension. He was a current smoker with a history of 2 packs of cigarettes every day for 45 years.

He confirmed progressively worsening chest pain. His dyspnea was estimated to be graded 1 mMRC. However, there was no history of wheeze, cough, anorexia, or weight loss.

The general physical examination was normal. On chest examination, he maintained saturation of 98% and showed symmetric chest wall expansion with normal vesicular breath sound. Lab investigations were within normal limits with no elevation of inflammatory biomarkers.

A Chest X-ray revealed opacity of the right upper lobe. A computed tomography (CT-scan) of the chest with contrast was performed and demonstrated a 6 × 3 cm, soft-tissue tumor mass, with irregular and speculated enhancements located between the right upper and the mild lobe. The right upper lobe bronchus was embedded in the mass which is, also invading the right pulmonary artery and the pericardium (Fig. 1).

Given his smoking history, a flexible bronchoscopy examination was performed and revealed that the lumen of the anterior bronchiole was occluded with an enlarged spur of the right upper main bronchus. Histopathologic examination of the biopsy sample revealed no malignant cells.

The patient then underwent a right upper lobectomy associated to a complete mediastinal and pulmonary lymph node dissection. Both the distal and proximal margins were negative on frozen section. The postoperative course was regular and the patient was discharged 3 days after surgery.

On macroscopic evaluation, the tumor nodule was well-circumscribed, smooth surfaced and completely occluding the bronchial lumen.

The postoperative pathology study showed bland-looking spindle cells arranged in fascicles with peripheral palisading of nuclei and slight myxoid background associated with formation of Verocay bodies. These cells showed a low mitotic activity. There was no evidence of tumor in lymph nodes. Immunohistochemical study revealed positive staining for S100 protein and negative staining for Pancytokeratin, epithelial membrane antigen, melan A, CD117, CD34, ALK and caldesmone.
Fig. 1. Computed tomography of the chest with contrast showing a right pulmonary mass with irregular enhancements causing an upper lobe consolidation (blue arrows pointing to endobronchial tumor mass and atelectasis).

Fig. 2. The microscopic slides illustrated:

(a) Well-circumscribed mass composed of uniform spindle cells arranged in palisading, with ovoid-to-round-shaped nuclei located in the eosinophilic collagen. No atypia or mitoses were identified.

(b) Immunohistochemical study: strongly and diffusely positive for S100 protein.

3. Discussion

Schwannomas are benign tumors that arise from peripheral, spinal, or cranial nerves [2]. They may occur anywhere on the body, but it is most commonly located on the flexor surfaces of the extremities, cerebellopontine angle, posterior mediatinum and spinal roots [3]. Primary neurogenic tumors are mostly associated with neurofibromatosis. However, primary endobronchial schwannoma, which is an exceedingly rare entity, can occur without associated neurofibromatosis as reported in our case.

In 1989, Feldhaus et al. described the first case of endobronchial schwannoma [4]. To our knowledge, there are approximately 50 case reports mostly in the Japanese literature and only scattered case reports from other countries [5].

Schwannomas can occur in any area of the tracheobronchial tree when they are located in the trachea or proximal bronchi they are called central tumors and peripheral when they cannot be reached by bronchoscopy. These tumors could have intraluminal or extraluminal extensions [6]. These endobronchial tumors can occur most often in the third decade of life, with no differences registered between gender [7]. Ethopathological mechanisms are still unknown. Nevertheless, some multiple sites of involvement suggest a possible underlying susceptibility to the development of these lesions.

The clinical course of endobronchial schwannoma depends on the site, the size and degree of bronchial obstruction [8]. Similar to our patient, symptoms include hemoptysis and dyspnea. However, fever, dry or productive cough and postobstructive pneumonia are not rare. As the symptoms of bronchial schwannoma are nonspecific, the diagnosis cannot be based on clinical presentation.

Radiological presentation of these lesions mainly included round or lobulated homogenous mass. Endobronchial nodule with partial or complete lumen obstruction was commonly associated with distal atelectasis or consolidation, as shown the CT of our patient.

Both clinical and radiological findings vary, and CT imaging cannot differentiate the nature of the tumor. The differential diagnosis of endobronchial tumors includes benign and malignant neoplasms such as hamartomas, carcinoids, fibrous histiocytomas, fibrous polyps, papillomas and leiomyomas [5].

The diagnosis of schwannoma is confirmed by histopathologic examination. It is characterized by the presence of typical Antoni A formation on hematoxylin-eosin stain and positive S100 protein on immunoperoxidase stain [6].

Bronchoscopy is often used in order to identify the location, the size and the degree of the bronchus lumen obstruction. Bronchoscopic biopsy will yield the diagnosis of proximal tumors. In this case, it also gives the opportunity to do curative therapeutic [9].

Treatment approaches for pulmonary schwannomas vary. The treatment of choice remains surgical resection. Our patient performed a right upper lobectomy, which is considered the standard treatment of these tumors. Surgical resection should be complete, to avoid local recurrence. In asymptomatic patients, the optimal monitoring and management strategy remains unknown. However, watchful waiting can be a therapeutic option [10].

Even schwannoma with malignant transformation is an exceedingly rare event, a long follow-up is needed for monitoring the tumor growth and reducing the risk of recurrence [11].
4. Conclusion

Endobronchial schwannoma is a benign and rare tumor with no specific clinical or radiological features. The optimal treatment is still until recently the surgical resection. This tumor is characterized by an optimistic prognosis with a rare risk of recurrence or a malignant transformation. However, a regular follow-up appointment is recommended.

Declaration of competing interest

Corresponding author: Imen TOUIL. Manuscript title: Endobranchial schwannoma in adult: a case report I certify that ALL of the following statements:

- The manuscript represents valid work; neither this manuscript nor one with substantially similar content under my authorship has been published or is being considered for publication elsewhere (except as described in the manuscript submission); and copies of any closely related manuscripts are enclosed in the manuscript submission;
- I agree to allow the corresponding author to serve as the primary correspondent with the editorial office and to review and sign off on the final proofs prior to publication; or, if I am the only author, I will be the corresponding author and agree to serve in the roles described above;
- Conception and planning of the work that led to the manuscript or acquisition, analysis and interpretation of the data, or both;
- Drafting and/or critical revision of the manuscript for important intellectual content;
- Approval of the final submitted version of the manuscript;
- I certify that I have participated sufficiently in the work to take public responsibility for the entire content of the manuscript.

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