Unusual presentation of massive intrathoracic schwannoma with concomitant pleural tuberculosis – case presentation and review of literature

Asociere neobișnuită de schwanom intratoracic masiv cu tuberculoză pleurală concomitentă – prezentare de caz și review al literaturii

Abstract

Schwannomas are the most common neurogenic tumors of the thorax, and may involve any thoracic nerve(1). We report the case of a 49-year-old male patient with a massive intrathoracic schwannoma. A thoracotomy resection of the tumor was performed. The presence of adhesions of the tumor to the chest wall pleura raised the suspicion of a malignancy. This article’s aim is to draw the attention to this infrequent pathology with concomitant tuberculosis, and to discuss different aspects regarding diagnosis and treatment of pleura schwannoma

Keywords: neurogenic tumor, pleural schwannoma, intrathoracic tumor

Rezumat

Schwannoamele sunt cele mai frecvente tumori neurogene localizate la nivelul toracului și pot afecta orice structură nervoasă de la acest nivel. Prezentăm cazul unui pacient de 43 de ani cu schvanom intratoracic masiv. S-a efectuat toracotomie cu rețeția tumorii. Aderența formăriilor la peretele toracic a ridicat suspiciunea unui proces proliferativ. Scopul articolului este să atragă atenția asupra acestei patologii rare asociate cu tuberculoză și să discute diferitele aspecte legate de diagnosticul și tratamentul în schwanomul pleural.

Cuvinte-cheie: tumoră neurogenă, schwanom pleural, tumoră intratoracică

Case Presentation

A 49-year-old male with no significant clinical history presented to the emergency department with a one-month history of productive cough with white sputum, haemoptysis and unexplained weight loss, associated with progressive shortness of breath on exertion, which had persisted despite courses of co-amoxiclav. On examination, his vital parameters (temperature, oxygen saturation, blood pressure, respiratory rate, pulse rate) were all within the normal range for a man of his age. Physical examination revealed dull percussion note on right mid chest wall with diminished breath sound and occasional expiratory wheezing at the auscultation of the chest, and syndrome (facial swelling, neck distension). Lymph nodes examination is unremarkable and the rest of the examination was unrevealing. Standard chest X-ray showed a large, well-defined mass in the the right hemithorax, around 18 cm in diameter, associated with displacement of the trachea to the opposite side (Figure 1). Initially, this was thought to be a bronchogenic cyst or a lymphoma process. Computed tomography (CT) scan of the thorax demonstrated a large (20x15x13 cm) well-circumscribed, heterogeneously enhancing rounded mass in the right hemithorax whose density was mostly similar to fluid, associated with a solid component in the uppermost part. The mediastinal structures, particularly the trachea and right bronchus, were noticed to be compressed and displaced to the left side by this lesion (Figure 2). Examination of the sputum smear samples to identify AFB was positive. Fine needle aspiration was performed, but the specimen was insufficient for diagnosis. The results from percutaneous biopsy confirmed a pleural schwannoma and eliminated a lymphoma process. Surgery was planned after ruling out an intraspinal component of tumor. Complete excision of the well-encapsulated mass was achieved through the right posterolateral thoracotomy (Figure 3A).
A 20-cm tumor was found in the right chest cavity, adhering closely to the parietal pleura and superior vena cava, which was severely dilated. The mass was found to be benign schwannoma without malignant components and the anatomopathological examination of parietal pleura retained it to be tubercular (Figure 3B). The postoperative course of the patient was uneventful. The respiratory and vascular symptoms and the facial edema resolved immediately after the surgery. The patient returned to his normal life and recovered gradually through regular chest physiotherapy. He has been followed-up for one year, with no evidence of recurrence. Antitubercular treatment according to the National Tuberculosis Control Program (NTP) standards in our country was started and he performed well, being declared cured and in our regular follow-up.

Discussion

The primary pleural schwannomas are usually benign(2), yet malignancy has been reported in some cases(3). They account for 1-2% of all thoracic tumors (3). Pleural schwannomas can occur at all ages, though they are uncommon in children. Both genders are equally affected, predominantly in the third or fourth decades of life(4,5).

Shin-ichi Takeda et al. analyzed the records of 146 patients with intrathoracic neurogenic tumors who were treated over the past 50 years. 20.5% of the neoplasms were malignant, occurring predominantly in the first 5 years of life. This study concluded that the age of patient seemed to be the most important clinical parameter for distinguishing between histological type and rate of malignancy for neurogenic tumors(6).

ClINICAL CASES

Intrathoracic neurogenic tumors occur predominantly (90%) in the mediastinum and about 10% originate in the peripheral nerve fibers from the intercostal nerves or pleural nerves(7,8). Schwannomas (also known as neurilemmomas or neurinomas) are highly vascular nerve sheath benign tumors that arise from the neural crest-derived Schwann cells(9).

The risk of malignancy in a nerve sheath tumor is very small (2-5%), frequently associated with von Recklinghausen syndrome (4% of cases), other neurofibromatoses or radiation exposure, when the risk increases to 10-20%(7,10,11).

The association of schwannoma with concomitant tuberculosis is a very rare entity. A literature review was performed via PubMed, using the MeSH terms ‘benign schwannoma’ and ‘tuberculosis’ and identified two similar case reports of intrathoracic schwannoma associated with tuberculosis(7,12). However, we found only one case of similar size to our observation, reported by David D. Odell et al(10).

Up to 80% of cases are asymptomatic, and diagnosis in young and middle-aged adults is generally fortuitous, frequently appearing as incidental radiological findings(9,12,13).

In our case, the presentation with superior vena cava syndrome is rare, making our patient atypical, reflecting clinical mutility and insidious evolution which can spread over several years, and explains the occurrence of the giant forms of this type of tumor. These symptoms are due to compression of neighboring structures(14).

A variety of diagnostic imaging modalities can be utilized to delineate pleural schwannomas. Generally, chest wall neurilemmomas are evaluated by radiologic examination for planning the surgical approach, including CT and magnetic resonance imaging (MRI).
CT scan remains the diagnostic imaging modality of choice for these neoplasms. CT scan can outline the size, number, and exact location of the lesions. CT scan can also demonstrate cystic and/or solid components of the tumor. Unenhanced CT demonstrates well-marginated soft-tissue mass with possible surrounding fat attenuation (“split fat” sign), may contain areas of low attenuation corresponding to fat or cystic degeneration, and calcification may be seen in 10% of tumors. Contrast-enhanced computed tomography (CECT) shows an homogeneous enhancement in small tumors and heterogeneous enhancement in larger tumors. Malignant pleural schwannomas have similar features on CT scan; however, they are usually associated with the presence of pleural nodules, pleural effusions, and metastatic pulmonary nodules.

In the case of our patient, the presence of dense adhesions and increased vascularity were in favor of malignant tumor component. This could have been secondary to the extension of inflammation response and fibrous reaction from the pleura tuberculous on to the capsule of the tumor and mimicked the features of malignancy.

MRI is the preferred method for the assessment of peripheral nerve sheath tumors. Vessels in neurinomas are usually prominent and their rich vascular supply is reflected in the often intense enhancement of these tumors on imaging studies. Pleural schwannomas are typically hyper-/isointense on T1-attenuated images and heterogeneously hyperintense on T2-attenuated images. Particularly, MRI should be performed in patients with suspected posterior mediastinal neurogenic tumors to exclude intra-spinal tumor extension. Thus, preoperative radiologic evaluation alone is enough without needle aspiration biopsy.

For giant shapes like our case, intra-thoracic schwannomas should be included in the differential diagnosis of posterior mediastinal masses. They include sympathetic ganglion tumours (neuroblastomas, ganglioneuroblastomas, ganglioneuromas) and paragangliomas (chemodectomas and pheochromocytomas), lymphadenopathy, enteric cysts, bronchogenic cysts, oesophageal tumours, aneurysms and paraspinal abscess.

In the absence of MRI, as in our observation, a preoperative percutaneous biopsy of the tumor, when feasible, is necessary in order to lead to the definitive diagnosis of benign tumor and to avoid overtreatment.

Microscopically, Antoni A and Antoni B areas are revealed in the majority of pleural schwannoma cases. Antoni A represents areas of hypercellularity with Verocay bodies. Antoni B areas of myxoid/hypocellular exhibit degenerating changes (i.e., cyst formation, haemorrhage, calcification, xanthomatous infiltration and hyalinisation). Immunohistochemically, pleural schwannomas typically stain diffusely and strongly positive for S100 protein. On the other hand, they stain negatively for CD-15, CD-30, CD-34, and pan-cytokeratin.

The standard care of management of pleural schwannomas is primarily surgical resection. In literature, the authors are unanimous that, compared to thoracotomy, VATS has better outcomes in terms of duration of surgical procedure, the amount of blood loss and length of drainage time. Complete resection can be achieved with low morbidity (smaller incisions, thereby resulting in less pain, fewer lung complications, shorter hospital stays, more rapid recovery, with return to normal activities) and less aesthetic damage.

A case series of 60 consecutive patients with a neurogenic tumor of the chest during 30-year experience concluded that VATS seems feasible as the approach for the thoracic neurogenic tumor since it is less invasive and provides an appropriate view for the operation.
A combined Chinese experience reports a thoracoscopic removal successfully of intrathoracic neurogenic tumors on average 3.5 cm in greatest diameter (21). However, the video-assisted thoracoscopic may be contraindicated in tumors larger than 6 cm (14,22).

David D. Odell et al. report a case of intrathoracic schwannoma measuring 27 cm in diameter and conclude that the clamshell incision, compared with the median sternotomy, offers excellent exposure of the pulmonary hilum, which is especially helpful in the setting of centrally located tumors (10).

In our case, we decided to perform an approach through a right posterior thoracotomy in the 6th intercostal space due to the size of the lesion and the possibility of an intraspinal component of the tumor, which necessitate a combined or staged approach, involving both neurosurgical and thoracic procedures.

Conclusion
This case report highlights the need to be aware of the potential coexistence of tuberculosis and intrathoracic schwannoma, hence the necessity of a systematic identification of an AFB on a sputum smear samples before any thoracic surgery in our country. Conversely, any positivity of the AFB test should not conclude for any intrathoracic mass to be a tuberculoma, without the confirmation by the examination of the tissue through a biopsy or surgical excision.

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Authors affiliation:
1. Teaching Hospital of Joseph Ravoahangy Andrianavalona Ampeliloha, Department of Surgery & Division of Thoracic Surgery, BP: 4150, Faculty of medicine of Tananarive CP: 101
2. Teaching Hospital of Joseph Ravoahangy Andrianavalona Ampeliloha, Anaesthesia reanimation units, BP: 4150, Faculty of medicine of Tananarive CP: 101
3. Teaching Hospital of Joseph Ravoahangy Andrianavalona Ampeliloha, Department of Surgery & Division of Pediatric Surgery, BP: 4150, Faculty of medicine of Tananarive CP: 101

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