Primary intrapulmonary benign schwannoma. A case with ultrastructural and immunohistochemical confirmation

X. Bosch, J. Ramírez, J. Font, J.A. Bombi, J. Ferrer, J. Vendrell, M. Ingelmo

ABSTRACT: Neurogenic pulmonary tumours are rare. Neurofibromas are the most common variety and tend to occur in patients with generalized neurofibromatosis, whereas schwannomas arise sporadically in patients who have no evidence of this disorder. A case of solitary benign schwannoma arising in a segmental bronchus is presented, and the published cases reviewed. Microscopically, the tumour had a spindle-shaped cell proliferation with palisade formation in several fields and low cellularity in other areas. The diagnosis was supported by positive immunostaining for S-100 protein and by ultrastructural examination which showed elongated cells surrounded by well-preserved basal lamina and numerous cytoplasmic processes also possessing a basal lamina.

Correspondence: Dr X. Bosch, Medicina Interna General, Unidad 1, Hospital Clinic i Provincial, Villarroel 170, 08036 Barcelona, Spain.

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Neurogenic tumours can arise anywhere in the thorax, but are most frequently found in the costovertebral angle [1-4]. Primary intrapulmonary tumours of neurogenic origin are extremely uncommon [1-3, 5, 6]. ASKANAZY et al. [2] first mentioned the presence of bronchial neurofibroma (NF) in generalized neurofibromatosis in 1914, and the first detailed description was published by RUBIN et al. [7] in 1940. Since then, only a few acceptable cases have been reported [1, 2, 6]. Histologically, the majority of the neurogenic tumours are NF, and schwannomas represent a distinct rarity [1, 5, 6].

Case report

A 72 yr old woman gave a two week history of a painful and swollen left calf. Past medical history was unremarkable.

Blood pressure was 140/80 mmHg, pulse rate 66min⁻¹, and temperature 37.2 °C. The left calf was moderately swollen, and warm, and Homans’ sign was positive. There were no other significant findings on physical examination. A full blood count and biochemical profile were normal. Contrast venography was consistent with thrombosis of left tibial and peroneal veins.

Chest X-ray showed a 5.5 by 3 cm sharply defined round mass in the anterior segment of the left upper lobe (LUL); no calcification or cavitation was observed. Computerized tomography (CT) confirmed the intrapulmonary location of the mass, showing a homogeneous pattern (fig. 1). Bronchoscopy revealed a small round tumour arising in a segmental branch of the LUL bronchus, causing 50% occlusion of its lumen. When touched, the mass bled easily and only one biopsy could be taken; this revealed only a non-caseating granuloma in the lamina propria.

Fig. 1. – Computerized tomography appearance of the mass confirming its intrapulmonary location and showing homogenous pattern.

At left posterolateral thoracotomy there were no enlarged nodes or pleural changes. A mass was felt in the anterior segment of the LUL. A pneumotomy was performed, and a round well-demarcated tumour was totally resected from healthy pulmonary parenchyma. The patient did well post-operatively. She was discharged on the eleventh post-operative day, and was asymptomatic six months following the operation.
Grossly the tumour was lobulated, encapsulated and pale-tan coloured. The consistency was firm and after slicing it showed a solid, white and partly fasciculated appearance. No haemorrhagic or necrotic areas were evident.

Microscopically the tumour was quite heterogeneous due to a variable cellular density. There was a spindle-shaped cell proliferation with palisade formation in several fields and low cellularity in other areas. The cells were wavy, with central and oval nuclei. There were neither mitotic figures nor atypical cells. No lung remnants were present, other than a few small cavities lined by simple flat epithelium in the periphery of the tumour (fig. 2).

The microscopic features were those of a benign schwannoma. This diagnosis was supported by positive immunostaining for S-100 protein (polyclonal antibody from Dako, Denmark) (fig. 3) and by electron microscopic examination, which showed elongated cells surrounded by well-preserved basal lamina. Numerous cytoplasmic processes also possessing a basal lamina and occasionally joined by desmosome-like junctions were frequently observed. Within the cytoplasm the cells had scattered microfilaments and scarce mitochondria, lysosomes and endoplasmic reticulum (fig. 4).

Discussion

Benign schwannomas or neurilemmomas (NL) are nerve-sheath tumours which arise wherever there are medullated nerves [9], spinal nerve roots being the most common primary location [9, 10]. They are very rare in bronchi or within the pulmonary parenchyma [6, 8]. Only 13 cases have been previously reported. Lane et al. [11] described in 1953 the first proven case of intrapulmonary NL, which was confirmed by tissue culture. On reviewing the reported cases in 1965, Bartley et al. [6] found that NL constituted one fourth of pulmonary neurogenic tumours; they found 7 reports of intrapulmonary or bronchial NL, but one of these [3] can be excluded because of its primary tracheal location [12]. Table 1 summarizes the reported proven cases of intrapulmonary and bronchial NL. We have not included 2 cases referred to by Silverman et al. [5] (those of Hochberg and of Gautam) because of erroneous references.

NL of the lung is a benign neoplasm that is usually detected during routine X-ray examination, most patients being asymptomatic [1, 6, 8]. The clinical course depends on the degree of bronchial obstruction and the tumour size attained [6, 13]. Symptoms are usually mild, consisting of dry or productive cough, chest pain, fever, and haemoptysis [2, 6]. Radiologically, the tumours appear as round, ovoid or lobulated, homogenous masses with a sharp outline, and occasionally with spotty calcification [6, 8]. When a large bronchus is compressed by the tumour atelectasis may be the only radiologic sign [6, 14]. The CT appearance of NL has been described as a well-circumscribed, homogeneous mass of soft tissue density [4], whereas inhomogeneity would be suggestive of malignancy. However, NL may have areas of inhomogeneity on CT due to haemorrhage or necrosis [15]. At bronchoscopy, the lesion may be seen as a polypoid mass bulging into the bronchial lumen or raising the bronchial mucosa, which may be then ulcerated [2, 6, 13].
The association between primary intrapulmonary NF and von Recklinghausen's disease is well known [1-3]; however, there was no evidence of this disease in all the cases of intrapulmonary NL recorded in table 1. Usually, NL are solitary tumours that arise sporadically in patients who have no evidence of a genetic predisposition. Nevertheless, if pigment changes in the skin are associated with a schwannoma, generalized neurofibromatosis should be suspected, as previously reported [9].

On gross pathologic examination, NL are usually quite small, well-encapsulated and of firm consistency. The cut surface is relatively homogeneous, and cystic zones may be present; necrosis, haemorrhage, and calcification are uncommon features [2, 6, 8, 9, 13, 16].

Histologically, a typical NL is sharply circumscribed by a thin fibrous capsule which is formed by compression of perineural tissue [6, 9]. Two types of tissue (Antoni A and B) are found. Antoni type A (cellular pattern) is formed of compactly arranged spindle cells with elongated nuclei disposed in parallel rows, creating a pattern of palisades. Admixed with this or in separate areas, the tumour has a less cellular Antoni B pattern with elongated cells arranged in irregular fashion and separated from one another by a matrix that stains poorly or not at all with haematoxylin and eosin and alcian blue stains [6, 8, 9, 13, 15, 17].

Clinically, the differential diagnosis of pulmonary NL is that of a pulmonary round focus. Histologically, spindle cell tumours must be ruled out, leiomyoma, fibroma and sclerosing haemangioma being the usual types in this location [1, 6, 9]. The presence of Antoni type A and B areas is very helpful, but it is preferable to demonstrate the neurogenic nature by immunohistochemical and ultrastructural studies [5, 18].

Identification of S-100 protein in both nucleus and cytoplasm of the tumour cells after using a polyclonal S-100 antiserum staining supports the schwannian origin of this neoplasm [18, 19].

Ultrastructural features of NL include the presence of abundant basal lamina surrounding the elongated cells, and numerous cytoplasmic processes, also with basal lamina, occasionally joined by desmosome-like structures [5, 9, 20].

Finally, the encapsulation of these tumours usually permits complete surgical enucleation, although, sometimes, resection of adjacent lung tissue may be indicated [2, 3]. In patients with intrabronchial tumours, the mass may be removed endoscopically [3, 6].

References


Table 1. – Reported cases of intrapulmonary and bronchial neurilemoma

<table>
<thead>
<tr>
<th>Author/Reference/Year</th>
<th>Age (yrs)</th>
<th>Sex</th>
<th>Symptoms</th>
<th>Location</th>
<th>Chest radiology</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lane et al. [11] 1953</td>
<td>16</td>
<td>F</td>
<td>Asymptomatic</td>
<td>RUL</td>
<td>Oval mass</td>
</tr>
<tr>
<td>Thoruskud [6] 1960</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Nakagawa [6, 14] 1961</td>
<td>32</td>
<td>F</td>
<td>Cough, fever and sputum</td>
<td>LLL</td>
<td>Atelectasis</td>
</tr>
<tr>
<td>Silverman et al. [5] 1976</td>
<td>63</td>
<td>M</td>
<td>Chest pain</td>
<td>RML</td>
<td>Round mass</td>
</tr>
<tr>
<td>Velthi et al. [8] 1982</td>
<td>73</td>
<td>F</td>
<td>Upper respiratory tract infection</td>
<td>RUL</td>
<td>Nodular density</td>
</tr>
<tr>
<td>Roviaro et al. [2] 1983</td>
<td>45</td>
<td>M</td>
<td>Asymptomatic</td>
<td>LLL</td>
<td>Round opacity</td>
</tr>
<tr>
<td>Roviaro et al. [2] 1983</td>
<td>47</td>
<td>F</td>
<td>Haemoptysis</td>
<td>LLL</td>
<td>Shadow</td>
</tr>
<tr>
<td>Roviaro et al. [2] 1983</td>
<td>50</td>
<td>M</td>
<td>Asymptomatic</td>
<td>LLL</td>
<td>Round mass</td>
</tr>
<tr>
<td>Bosch et al. 1989</td>
<td>72</td>
<td>F</td>
<td>Asymptomatic</td>
<td>LUL</td>
<td>Round mass</td>
</tr>
</tbody>
</table>

R: Right; L: Left; RUL: right upper lobe; LUL: left upper lobe; LLL: left lower lobe; RML: right middle lobe.
11. Lane N, Murray MR, Fraser GC. - Neurilemoma of the lung confirmed by tissue culture. *Cancer,* 1953, 6, 780–785.


RÉSUMÉ: Les tumeurs pulmonaires neurogènes sont rares. Les neurofibromes en sont la variété la plus commune et tendent à se développer chez des patients atteints de neurofibromatose généralisée, alors que les schwannomes apparaissent de façon sporadique chez des patients sans aucun signe de cette maladie. Un cas de schwannome bénin isolé dans une branche segmentaire, est exposé avec une revue des cas publiés. À l’examen microscopique, l’on a noté dans plusieurs champs une prolifération de cellules fusiformes avec des formations palissadiques; la cellularité était faible dans d’autres zones. Le diagnostic est confirmé par une immunocoloration positive pour la protéine S-100. L’examen au microscope électronique montre des cellules allongées entourées par une lamina basa bien préservée et par de nombreux processus cytoplasmiques possédant également une lamina basa.