Case 10514

Melanocytic schwannoma of the phrenic nerve
Published on 03.12.2012

DOI: 10.1594/EURORAD/CASE.10514
ISSN: 1563-4086
Section: Chest imaging
Area of Interest: Thorax
Procedure: Diagnostic procedure
Procedure: Instrumentation
Imaging Technique: Conventional radiography
Imaging Technique: PET-CT
Imaging Technique: Experimental
Imaging Technique: CT
Special Focus: Neoplasia Case Type: Clinical Cases
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Patient: 49 years, female

Clinical History:
A non-smoking 49-year-old woman with no relevant medical history presented with lower left chest pain for 6 months and dry cough. On clinical examination, left basal hypoventilation was noted. Laboratory findings were normal.

Imaging Findings:
Chest radiograph showed a left basal thoracic opacity with a broad pleural base suggesting a mass of pleural origin versus pleural effusion (Fig. 1). Computed tomography (CT) showed a heterogeneous soft tissue lesion measuring 20 cm in the axial plane, involving the left basal pleura and extending into the oblique fissure with central calcifications and multiple fluid-density loculated cavities (Fig. 2).
Positron emission tomography-CT (PET-CT) showed a hypermetabolic mass adjacent to the left lower lobe (standardized uptake value max. 14) (Fig 3). No malignant cells were found on bronchoscopy.

Discussion:
Surgical excision with tumourectomy was performed in our patient. A voluminous mass adjacent to the left diaphragm was confirmed, with involvement of the phrenic nerve (sacrificed during surgery) and the diaphragm itself. The definitive histology showed a melanocytic schwannoma with positive S100 protein marker (Fig. 4).

Intrathoracic neurogenic tumours are common but most often affect the intercostal nerves or the sympathetic chain. Their most common location is the posterior mediastinum. The phrenic nerve is not a frequent location with only 3 cases reported in the literature [1].
Tumours arising from nerve sheaths (schwannomas, neurofibromas, and malignant peripheral nerve sheath tumours- MPNST) are differentiated from those arising from nerve cells - neuromas [1]. Schwannomas are benign nerve tumours composed of Schwann cells.
Melanocytic schwannoma, however, is a rare and potentially malignant neoplasm. This tumour contains epithelioid cells with increased melanin content [2]. Apart from the presence of melanin, it has other unusual characteristics that distinguish it from schwannoma which include also psammoma bodies [3] or positive S100 protein marker as in our case. Pathologically, one important differential diagnosis of melanocytic schwannoma would be metastatic or primary melanoma. Melanocytic schwannoma shows considerably less cytologic pleomorphism and mitoses than
one would expect in melanoma.

The clinical presentation of melanocytic schwannoma is not specific and depends on its location. Patients with thoracic involvement, like in our case, may present with cough, hoarseness, weight loss, and less commonly with diaphragmatic paralysis or diaphragmatic eventration.

On imaging studies, schwannomas appear usually as homogeneous and well-circumscribed lesions of soft-tissue density on non-enhanced CT [4, 5]. After contrast administration, the enhancement is variable and can be either homogeneous or heterogeneous, especially in larger lesions with zones of necrosis. Imaging modalities such as CT or MRI are inevitable to define the exact location of the lesion and to exclude a vascular origin [6]. Nevertheless, histology remains the gold standard for final diagnosis and allows exclusion of malignant lesions.

The prognosis of melanocytic schwannoma implicates long-term follow-up, even after radical excision. The tumour can be aggressive with local recurrences occurring years after surgery. There is also a potential of malignant degeneration [7].

The take home message is that despite neurogenic tumours being frequent paraspinal lesions, they may appear in unusual locations outside of the posterior mediastinum and mimic other lesions. The particular case of melanocytic schwannoma underlies the importance of histological analysis in the final diagnosis.

**Differential Diagnosis List:** Melanocytic schwannoma of the phrenic nerve, Localized fibrous tumour of the pleura, Metastasis, Lymphoma, MPNST (malignant peripheral nerve sheath tumour), Neurofibroma, Schwannoma, Sarcoma, Melanoma

**Final Diagnosis:** Melanocytic schwannoma of the phrenic nerve

**References:**


Description: Left basal thoracic opacity with a broad pleural base suggesting a mass of pleural origin or pleural effusion. Origin: Geneva University Hospital
Description: Heterogeneous soft tissue density lesion at the left lung base involving the basal pleura with central calcifications and multiple fluid-density cavities. Origin: Geneva University Hospital
Description: The lesion is extending into the oblique fissure. Origin: Geneva University Hospital
Description: Hypermetabolic mass adjacent to the left lower lobe. Origin: Geneva University Hospital
Figure 4

a

Description: Macroscopic aspect of one fragment of the lesion after fixation. Origin: Geneva University Hospital

b

Description: Tumour cells showing nuclei and lighter colored nucleoli. Origin: Geneva University Hospital