Primary intrapulmonary malignant peripheral nerve sheath tumor mimicking lung cancer

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Malignant peripheral nerve sheath tumors (MPNSTs) are sarcomas originating from the cells constituting the nerve sheaths such as Schwann cells, perineural cells or fibroblasts. They represent approximately 5-10% of all soft tissue sarcomas. They have been rarely observed in the lung. We describe a rare case of primary lung MPNST in an elderly male patient, in which surgical approach has obtained a good control of the disease. Immuno-histochemical and molecular analyses have been required on the surgical specimen due to inadequate possibility of recognition through morphology alone.

Malignant peripheral nerve sheath tumor; schwannoma; lung cancer
Figure 1. A. Chest CT showing the left lower lobe nodule; B. Surgical specimen.

Figure 2. A. Proliferation of mesenchymal neoplastic spindle cells densely packed, arranged in short bundles (H&E, ×20); B. Negative immunostaining for low molecular weight keratin (panCK, ×40); C. Strong and uniform positivity for vimentine (IHC, ×40); D. Negative immunostaining for desmina (IHC, ×40); E. Cytoplasmatic immunostaining for S-100 (IHC, ×40); F. Low positive immunostaining for EMA (IHC, ×40); G. Representative samples of break-apart FISH assay for SYT rearrangement. Our sample shows paired signals (orange/green) consistent with the presence of the intact SYT gene while positive tumor cells should exhibit SYT disruption associated with increased number of paired and unpaired signals, represented by one paired signal with multiple orange and green unpaired signals.
sheath origin, only one case of benign schwannoma was observed in the lung (5). The incidence of MPNST in the general population is 0.001%; however, it increases to 5-42% in patients with type 1 neurofibromatosis (NF1) (2). Clinical symptoms are usually similar to those observed in bronchogenic carcinomas, presenting with chest pain and cough generally occurring in the middle age with a slight predominance in males (3).

Reportedly, 5-year survival rates vary from 15% to 40% and patients with NF1 have generally a poorer prognosis. Other adverse prognosticators are tumor size greater than 5 cm, mitotic rate greater than 20×10 HPFs, central location and incomplete resection. MPNSTs are associated with high local recurrence rate. Moreover, distant metastases develop in more than one half of patients. Lungs, bones, pleura and liver are commonly involved with diffusion via the meningeal route also being possible. Cytogenetic studies have shown complex clonal abnormalities in most cases (3).

In the differential diagnosis, pulmonary metastases from extrathoracic mesenchymal tumors should be ruled out. However, the distinction from other primary sarcomas of the lung is still very difficult. In particular, the MPNSTs have to be distinguished from other mesenchymal malignant neoplasms (such as synovial sarcoma and leiomyosarcoma) especially in cases, like ours, when aberrant EMA expression is observed. In this context, the resort of immunohistochemistry is not helpful and molecular characterization is needed for a correct diagnosis (6). In order to obtain local control, malignant mesenchymal tumors could be approached surgically with satisfactory results as in our patient.

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Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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References