A rare case of synchronous malignant thoracic tumors*

Um caso raro de tumores torácicos malignos sincrônicos

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Abstract
Malignant neurogenic mediastinal tumors in adults are uncommon and extremely aggressive. We report the case of a 61-year-old male patient with the simultaneous occurrence of malignant mediastinal schwannoma and bronchioloalveolar carcinoma. Although bronchioloalveolar carcinoma is present in 4-7% of the resected synchronous thoracic tumors, this association has never been reported in the literature. However, it is a common finding in patients presenting apparently inflammatory infiltrates and ground-glass opacities, as in the case presented here.

Keywords: Mediastinal neoplasms; Nerve sheath neoplasms; Neurilemmoma; Neoplasms, multiple primary; Adenocarcinoma, bronchiolo-alveolar.

Introduction
Neurogenic tumors are the most common mediastinal tumors, accounting for 20-35% of all of the mediastinal neoplasms.\(^1\)\(^,\)\(^2\) Neurogenic tumors are also responsible for approximately 75% of all lesions in the posterior mediastinum, although they are typically benign and asymptomatic. When symptoms are present, they should raise the suspicion of a malignant lesion.\(^3\)

Neurogenic mediastinal tumors can originate in any neural structure within the chest and are classified according to their origin: those originating in the peripheral nerve sheaths, those originating in the sympathetic nervous system and those originating in the parasympathetic nervous system. Sympathetic tumors are classified as neuroblastomas, ganglioneuroblastomas or ganglieneuromas. Parasympathetic tumors are uncommon and include functioning paragangliomas (pheochromocytomas) and nonsecreting chemodectomas.\(^1\)

Of all of the neurogenic mediastinal tumors, 40-65% originate from the peripheral nerve sheath. Schwannomas (neurilemmomas) and neurofibromas, both benign lesions, account for over 95% of the tumors in this group. Malignant peripheral nerve sheath tumors (malignant schwannomas) are extremely aggressive and account for the remaining 2-40%.\(^2\)

Case report
A 61-year-old male patient sought treatment in the emergency room of our institution reporting subcapular pain for two months. The

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Financial support: None.
patient reported that, in the beginning, the pain was tolerable when controlled with common analgesics. However, it became increasingly intense. There were no symptoms other than pain, and the patient reported no cough, hoarseness, fever, chills, night sweats, weight loss, hemoptysis, dyspnea or exposure to TB. He was a 30-pack-year smoker and a social drinker. The patient stated that he had no family history of neoplasms.

A physical examination revealed good general health and nutritional status, and vital signs were normal. The patient presented no lymph node enlargement. Upon pulmonary auscultation, there were fine inspiratory rales throughout both lung bases. The cardiac auscultation was normal. Examination of the abdomen and lower limbs revealed no alterations. The results of the blood workup and biochemical analyses were within the limits of normality. A CT scan of the chest (Figure 1a) revealed a well-defined mass, with regular borders, in the right costovertebral region, measuring 6 × 4.3 cm. Adjacent to the mass, we observed pulmonary infiltrates, apparently inflammatory and nonspecific (Figure 1b).

A right posterolateral thoracotomy was performed. We observed a large unencapsulated mass, lobulated and densely adhered to the costovertebral region, extending from T7 to

![Figure 1](image1.png)

**Figure 1** - In a), well-defined mass, with regular borders, located in the right costovertebral region, without radiological signs of invasion of the spinal canal or adjacent structures. In b), area of ground-glass infiltrate (arrow) in the sixth segment of the right lower lobe, adjacent to the mediastinal mass.

![Figure 2](image2.png)

**Figure 2** - In a), high-grade spindle-cell sarcoma. We observe severe nuclear atypia. (H&E; magnification, ×400). In b), nonmucinous bronchioloalveolar carcinoma. Note malignant epithelial cells (arrows) around alveolar walls (H&E; magnification, ×400).
Malignant neurogenic mediastinal tumors are extremely aggressive and associated with a low five-year survival rate. Local invasion, hematogenous metastases and pulmonary metastases frequently occur. Local recurrence and metastases are usual. Findings suggestive of malignancy on CT scans are as follows: low density areas; compression of adjacent structures; pleural abnormalities, such as pleural effusion or pleural nodules; and metastatic pulmonary nodules. Erosion of bone structures and pain are not uncommon findings and also suggest malignancy. Radical surgical resection with wide margins is the treatment of choice. When complete resection is not feasible, a simple excision without wide margins or a subtotal excision followed by radiotherapy in high doses are possible alternatives. Adjuvant radiotherapy and chemotherapy do not seem to increase survival but are useful in the treatment of the metastatic disease. Surgical cure is rarely possible.

The simultaneous occurrence of a malignant mediastinal schwannoma and bronchioloalveolar carcinoma is unprecedented. Resection of multiple thoracic neoplasms accounts for approximately 4% of all lung cancer resections, and most cases occur between the sixth and seventh decade of life. The genesis of synchronous tumors can be attributed to pathogenic factors intrinsic to the individual or can be related to a random phenomenon. According to two authors, synchronous thoracic tumors are defined as those that are diagnosed simultaneously with the index tumor, are separated from the index tumor by areas of healthy lung parenchyma and do not share lymphatic drainage with the index tumor. Histopathological characteristics, morphology, location, vascular invasion and immunohistochemical analysis should also be taken into consideration in the differentiation of the tumors. Different histology in clearly distinct neoplasms is pathognomonic of the primary nature of the lesions. However, this occurs in only 10–15% of the cases.

Areas of ground-glass opacity on tomography scans of the chest are unspecific findings and may represent various conditions, such as pulmonary edema, alveolar proteinosis, alveolitis, interstitial pneumonitis and neoplasms. A diagnosis of neoplasia in ground-glass pulmonary infiltrates, as in this case, is common. One group
of authors evaluated 20 cases of patients with ground-glass infiltrates submitted to surgical resection. Of those, 50% were diagnosed with bronchioalveolar carcinoma and 10% were diagnosed with adenocarcinomas. In addition, 25% were diagnosed with atypical adenomatous hyperplasia, which is considered the precursor lesion of bronchioalveolar carcinoma.

Although it is the most common malignant neoplasia of the peripheral nervous system, malignant schwannoma is still one of the least studied sarcomas. The five-year survival rate is low and is negatively affected by the size of the lesion, incomplete resection and concomitance with von Recklinghausen disease. Complete resection is usually impossible. Adjuvant radiotherapy and chemotherapy can be useful in the treatment of the metastatic disease. The lesion is rare as are its symptoms. Therefore, a high level of clinical suspicion is recommended in order to correctly diagnose this rare neoplasia. It is likely that the combination of malignant schwannoma and bronchioalveolar carcinoma was a random phenomenon and did not influence the outcome of the case, which was a consequence of the aggressive nature of the mediastinal lesion. However, the presence of neoplasia should always be suspected in patients presenting pulmonary infiltrates and localized ground-glass opacities that do not disappear or grow. In such cases, an aggressive approach should be adopted, with early biopsy and histopathological analysis.

References


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