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An Atypical Neurofibroma in the Posterior Mediastinum: A Case Report

Posterior mediastinum yerleşimli atipik nörofibroma: Olgu sunumu

Summary
Nerve sheath tumors such as neurofibroma, schwannoma, and perineurioma are relatively uncommon lesions that sometimes constitute an interesting diagnostic and therapeutic problem in the clinical practice.
A 27-year-old male patient was operated for a mass located in the left posterior mediastinum. The mass was resected by a thoracotomy, and a diagnosis of atypical neurofibroma was revealed histopathologically. He has been well without any problems in his 6 months postoperative period.
Intrathoracic neurogenic tumors should be resected surgically due to the possibility of malignancy. Complete resection can be performed safely by a thoracotomy approach and is necessary for achieving a cure.

Key words: Mediastinum; Mediastinal Neoplasms; Nerve Sheath Neoplasms; Neurofibroma; and Thoracic Surgery

Özet
Sinir kılıfı tümörleri içerisinde değerlendirilen nörofibroma, schwannoma ve perinöroma nispeten nadir lezyonlardır ve klinikte bazen ilginç tanı ve tedaviye ait problemlere neden olmaktadır.
27 yaşındaki erkek olgu sol posterior mediastinum yerleşimli kitle nedeniyle opere edildi. Kitle torakotomi ile çıkarıldı ve histopatolojik inceleme ile atipik nörofibroma tanısı konuldu. Olgu 6 aylık takip dönemi sonunda asemptomatiktir.
İntratorasik nörojenik tümörler malignite ihtimali nedeniyle opere edilmelidirler. Tam rezeksiyon torakotomi ile güvenle yapılabilir ve kür elde edilmesi için gereklidir.

Anahtar kelimeler: Mediastinum; Mediastinal Tümörler; Sinir Kılıfı Tümörleri; Nörofibroma; ve Göğüs Cerrahisi

INTRODUCTION

Intrathoracic neurogenic tumors are uncommon neoplasms arising from the intercostal and sympathetic nerves. They are found almost exclusively in the posterior mediastinum around the paravertebral area (1). These tumors are usually benign causing no clinical symptoms at presentation, and are often detected incidentally on chest radiographs. At present, because of reasons such as the uncertainty of preoperative diagnosis, the increasing size of the tumor, and the possibility of malignancy, early surgical excision is considered to be the most acceptable strategy (2). Benign neurogenic tumors rarely recur after complete resection whereas malignant neurogenic tumors have poor prognosis. The purpose of this paper is to report a case of neurogenic tumor due to its extreme rareness.

Case report

A 27-year-old male patient was referred to our clinic because of an unusual appearance observed on the chest radiography taken on one of his routine check-ups. The results of his physical examination and laboratory tests were

normal. The chest radiography showed a mass like lesion in the left apical zone (Fig. 1). In the thoracic computed tomography; a round, homogeneous mass with well-defined borders, measuring 5 x 5 cm, was identified in the left superior posterior mediastinum. A left axillary thoracotomy revealed a brown and encapsulated solid mass. The mass was resected totally including its neurogenic pedicle. It weighed 65 g and measured 5 x 5.5 x 4.5 cm3 (Fig. 2).

Histopathologic examination was consistent with an atypical neurofibroma. Low-power microscopy showed a randomly arranged, vesicular growth pattern composing of spindle cells. The nuclei had a wavy pattern including a few atypical cells and a characteristically neural appearance. The cells were mixed with a fibromyxoid stroma and there was no increase in mitotic activity. Immunohistochemical staining (S100) confirmed the diagnosis (Fig. 3).

The patient was discharged following his recovery on the 6th postoperative day. No recurrence was observed radiologically during the 6 months follow-up period.

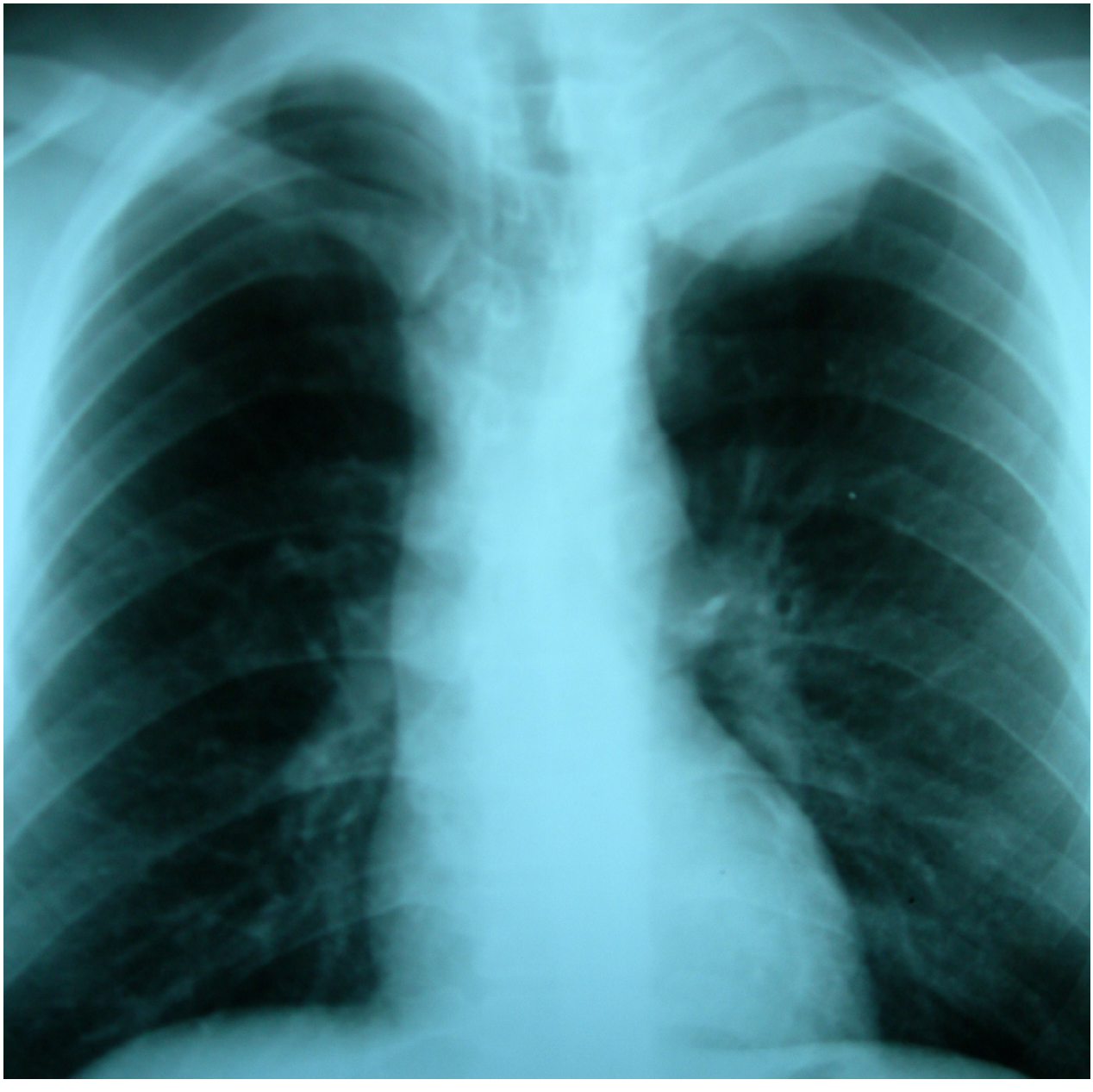

Figure 1. Chest X-ray image revealing a mass located at the left apical zone.

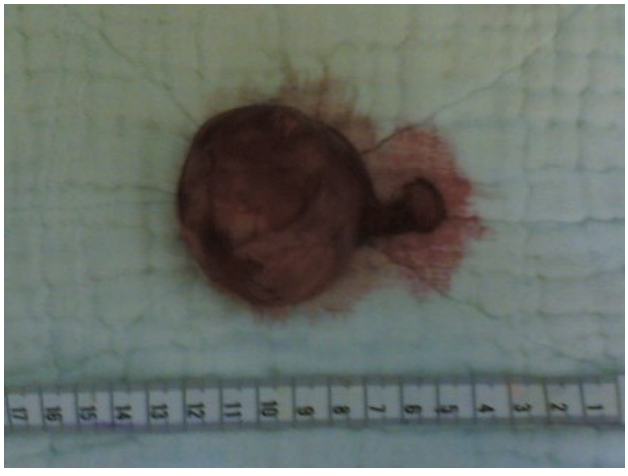

Figure 2. The macroscopic view of the resected specimen

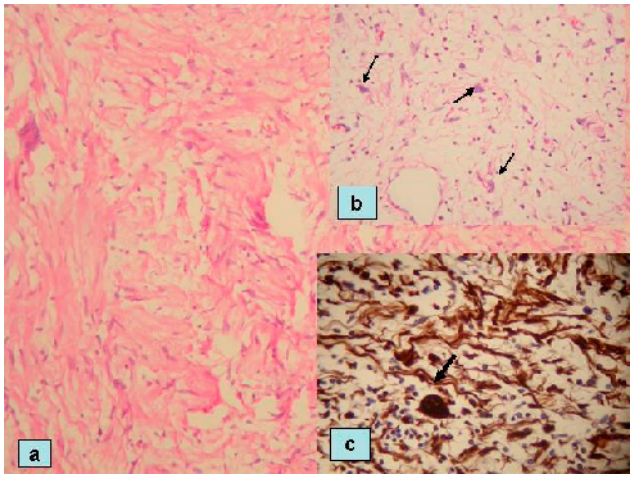

Figure 3. Histological examination revealed large, hypertrophic nerves that consisted mainly of spindle-shaped fibroblasts (a) (Hematoxylin-Eosin stain, original magnification x 100). The tumor cells occasionally included atypical cells that had large hyperchromatic, irregular nuclei (b) (Thin arrow) (Hematoxylin-Eosin stain, original magnification x 200), and were arranged in a distinct lamellar or fibrillar pattern. Mitoses were mostly absent. The pleomorphic cells expressed S-100 protein (c) (Thick arrow) and negative for p53 and MIB-1.

DISCUSSION

Intrathoracic neurogenic tumors have a variety of clinical and histological features and the majority of these tumors are asymptomatic. However, as they become larger in size, they may produce symptoms related to bone erosion, spinal cord involvement, and local compression on adjacent organs (2). Chest or back pain, coughing, dyspnea, wheezing, dysphagia, Horner's syndrome, arm paresthesia, and obstruction of the superior vena cava are among these symptoms. The patient presented in our case report had worked in the food sector and had no such complaints up to the time. The diagnosis was made based on the left apical opacity observed on the chest radiography taken on one of the carrier screenings carried out routinely each year.

In a retrospective study by Shrivastava et al., 106 patients underwent surgical treatment for a mediastinal mass and among 5 (4.7%) of the patients neurofibroma was diagnosed (3). Neurofibroma which is classified within benign nerve sheath tumors which constitute a subgroup of neurogenic tumors, is more common in adults than children, has a peak age of 20-40, shows no sex predilection and does not have any known serum tumor markers. Neurofibromas show degenerative changes occasionally and, at localizations where they occur as isolated neoplasms or as multiple thoracic manifestations of neurofibromatosis, they may exhibit malignant degeneration rarely but more often than Schwannomas (4). Neurofibromatosis is seen in 14-30% of the patients with mediastinal neurofibromas and most of these cases present at a younger age and is at increased risk for malignant transformation. The malignancy rates are reported to reach 30% in these patients (5). Consequently, it is advised that those cases which have been diagnosed as neurofibroma histopathologically, should be screened for Neurofibromatosis type 1 (von Recklinghausen disease). Neurofibromatosis type 1 is an uncommon autosomal dominant hereditary syndrome and is characterized with multiple light brown (café-au-lait) spots, optic gliomas, bone lesions, Lisch nodules (benign melanotic iris hamartomas) and neurofibromas widespread throughout the body. These neurofibromas may origin from vagus, phrenic, recurrent laryngeal or intercostal nerves, grow inside the mediastinum or thoracic wall and may reach up to big sizes (6). Unilateral apical neurofibromas as seen in our case are characteristical for Neurofibromatosis type 1 and may show aggressive behaviour. The reported case has been investigated in terms of Neurofibromatosis type 1 and no such diagnostic criteria have been found.

Essentially, the determination of the histopathological tissue diagnosis of the mediastinal masses in the preoperative period will specify the treatment method to be chosen. Such modalities include invasive ones such as anterior mediastinotomy, or minimal invasive ones such as transthoracic fine-needle aspiration guided by computed tomography or ultrasonography and transbronchial needle aspiration biopsy. However, in cases like ours where the masses are apically localized, or located adjacent to important

neurovascular structures or carry a risk of injury when exposed through the mentioned techniques, surgical excision by an exploratory thoracotomy will be definitely diagnostic and therapeutic. As it is known that the frequency of malignant tumors among intrathoracic neurogenic tumors is around 4-13% (2), an immediate complete surgical resection would provide more benefit than to conservatively follow the patients. When the invasive and aggressive nature of this tumor is considered; the diagnosis of atypical neurofibroma that we have derived from an asymptomatic case which we have operated on with no preoperative tissue diagnosis, seems to support the forementioned algorithm.

Surgical resection is considered as the treatment of choice for such tumors and thoracotomy has been the traditional surgical approach (7). Thoracotomy provides both surgical convenience and allows a complete resection with easy access and better visualization. This is a safe procedure with a very low mortality rate and an acceptable morbidity. However, the resection of tumors located in the apex of the chest cavity and in close vicinity to great vessels, brachial plexus, and the stellate ganglion, carries increased risk as for neurovascular injury and Horner's syndrome. Yet, avoiding blunt digital dissection without visual control and the careful dissection of the tumor from the surrounding tissues during the surgery, will considerably reduce the possible complication rates. The general rule that complete resection which is one of the general principles of tumor surgery is important to achieve satisfactory long term survival, is also valid for these kind of tumors. Hereby, the complete resection of the tumor and avoiding complications bear utmost importance.

As a conclusion, posterior mediastinal masses should be evaluated for thoracic surgery when diagnosed, and should be operated in order to ensure adequate complete excision and to provide histological diagnosis. Thoracotomy is a safe surgical approach with low complication risk, and allows the surgical team to easily evaluate the relationship of the tumor with the surrounding tissues.

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