

Pulmonary Atelectasia Caused by a Giant Cellular Schwannoma at the Dorsal Spine: Original Image

Dorsal Omurgadaki Dev Sellüler Schwannom'un Neden Olduğu Pulmoner Atelektazi

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ABSTRACT Cellular schwannoma (CS), which is a well-recognized variant of benign schwannoma, is often misdiagnosed as sarcoma. We report a case of giant CS, a rare benign nerve-sheath tumor in a 70 year-old woman. It presented as a voluminous lesion in the dorsal paraspinal region that caused left pulmonary atelectasia. These features, in association with the microscopic aspects of a hypercellular, pleomorphic neoplasm may lead to a false impression of a malignant tumor. In the treatment of the tumor, total removal of the lesion is considered curative and adjuvant chemotherapy or radiation therapy is not worth consideration. Therefore, it is important to have an accurate examination to confirm the benign nature of this tumor thus avoiding unnecessary therapy and its complications.

Key Words: Immunohistochemistry; nerve sheath neoplasms; neurilemmoma

ÖZET Selim schwannom'un iyi ayırt edilmiş varyantı olan sellüler schwannom (SS) sıklıkla sarkom olarak yanlış tanı almaktadır. Bu makale, 70 yaşındaki kadın hastada nadir görülen selim seyirli sinir kılıfı tümörü olan dev SŞ olgusunu sunmaktayız. Dorsal paraspinal bölgede oldukça geniş hacmiyle tanınan lezyon sol pulmoner atelektaziye neden olmuştur. Bu özelliklerinin mikroskopik olarak hipersellüler ve pleomorfik tümör görünümüyle birlikteliği, malign tümör olarak yanlış tanınmasına neden olabilir. Tümörün tedavisinde, lezyonun tümüyle çıkarılması tam olarak şifa sağlamakta ve cerrahi tedavi sonrası uygulanacak kemoterapi veya radyasyon tedavisinin iyileştirici etkisi bulunmamaktadır. Bu nedenle, tümörün selim doğasını doğrulamak için kesin değerlendirme yapılması önemlidir böylece gereksiz tedavi ve bu tedavinin komplikasyonları önlenir.

Anahtar Kelimeler: İmmünohistokimya; sinir kılıfı tümörleri; nörilemmoma

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Cellular schwannoma (CS) was first described by Woodruff et al in 1981 and has a female predominance, a median age of 55 years (range 17-79 years) and most are located in the mediastinum and retroperitoneum.^{1,2} CS accounts for approximately 5% of the benign peripheral nerve-sheath tumors (MPNSTs).³ This tumour is clinically unspecified and in most cases the symptoms are due to the compression of adjacent structures when its localization is retroperitoneal. With a varied presentation and a difficult preoperative diagnosis, schwannoma accounts for only a small percentage of retroperitoneal tumors (only 0.5-1.2%). Its diagnosis is quite

often confirmed by anatomopathological study afterwards.⁴⁻⁶ We report a case of this unusually large retroperitoneal pathology with uncommon presentation in a female patient.

A 70-year-old woman had a five years history of mild left chest pain. She had started to complain about respiratory distress two weeks ago. Chest X-ray showed left pulmonary atelectasia. An enhanced computed tomography (CT) scan revealed a $16.2 \times 12.7 \times 10.5$ cm heterogeneously enhancing retroperitoneal mass in the left paraspinal region, extending into the left T9-10 neural foramen. The patient was admitted to our clinic. Magnetic resonance imaging (MRI) confirmed the presence of the voluminous T9-10 left paravertebral mass. The neural foramen was expanded, the aorta was shifted laterally, the spleen was displaced anterolaterally and the diaphragm was elevated by the tumor which caused left pulmonary atelectasia (Figure 1). The neurological examination was normal. Café au lait spots, axillary freckling, or Lisch nodules were not observed. The patient displayed no familial history of neurofibromatosis.

The patient underwent a surgical procedure for total removal of the dorsal mass by retroperitoneal approach via a thoracolumbar incision in collaboration with the thoracic surgeon. The patient was placed at the right lateral decubitus and by retroperitoneal approach the paravertebral mass was radically removed en bloc. The tumor was solid and partially encapsulated; the anterior, posterior and both lateral portions revealed a well-defined capsule excluding the superior part (at this point, there was adhesion to the surrounding diaphragm). Thus, diaphragm repair was necessary in this case. Then, whole intraspinal tumor was removed using operating microscope. Despite the requirement of dural repair, resection of the affected nerve root was not necessary in this specific case for complete removal of the tumor.

The tumor was partially encapsulated except for an area that measured 2×1 cm. It weighed 984 g and measured $16 \times 13 \times 11$ cm (Figure 2). The cut surface was firm, yellow-white in color, including irregular hemorrhagic cystic areas. Histologically, specimens from individual tumors exhibited identical characteristics. This lesion showed compact

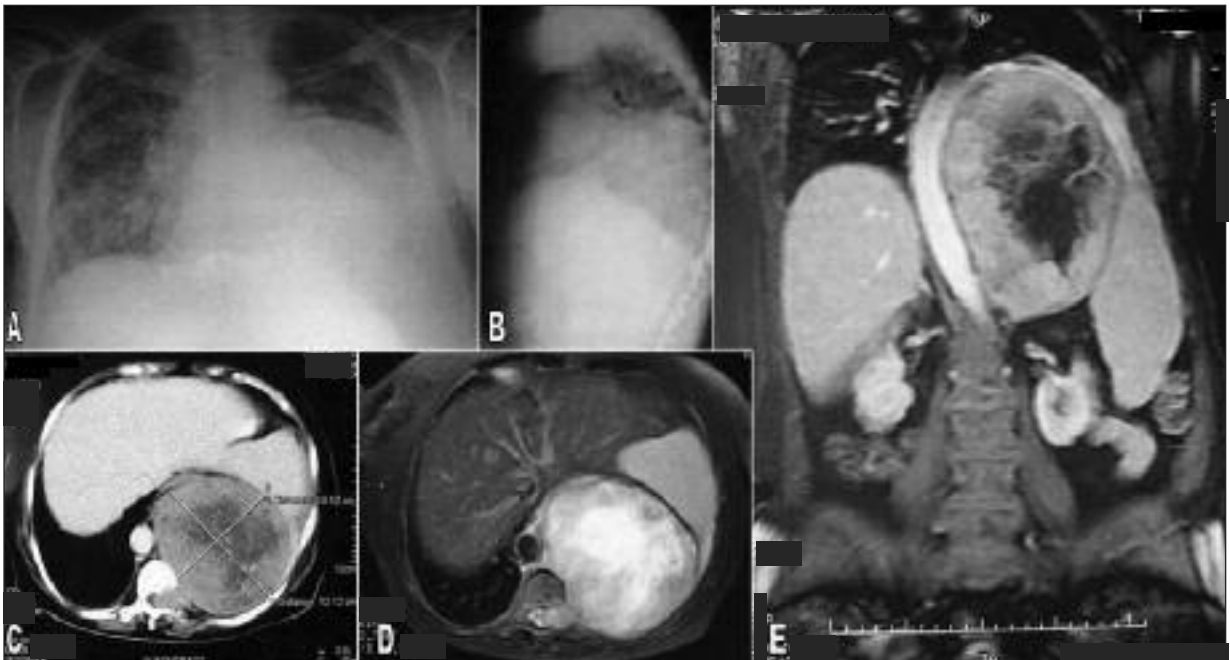


FIGURE 1: Anteroposterior (A) and lateral (B) chest radiographs, (C), (D) Preoperative axial CT scan and T2- weighted MRI images demonstrating a lesion expanding to the neural foramen, occupying the entire spinal canal from T9-10 and invading the retroperitoneum (E) Gadolinium-enhanced T1-weighted coronal thoracoabdominal MRI image demonstrating a large heterogeneously enhancing mass elevating the diaphragm and causing left pulmonary atelectasia.

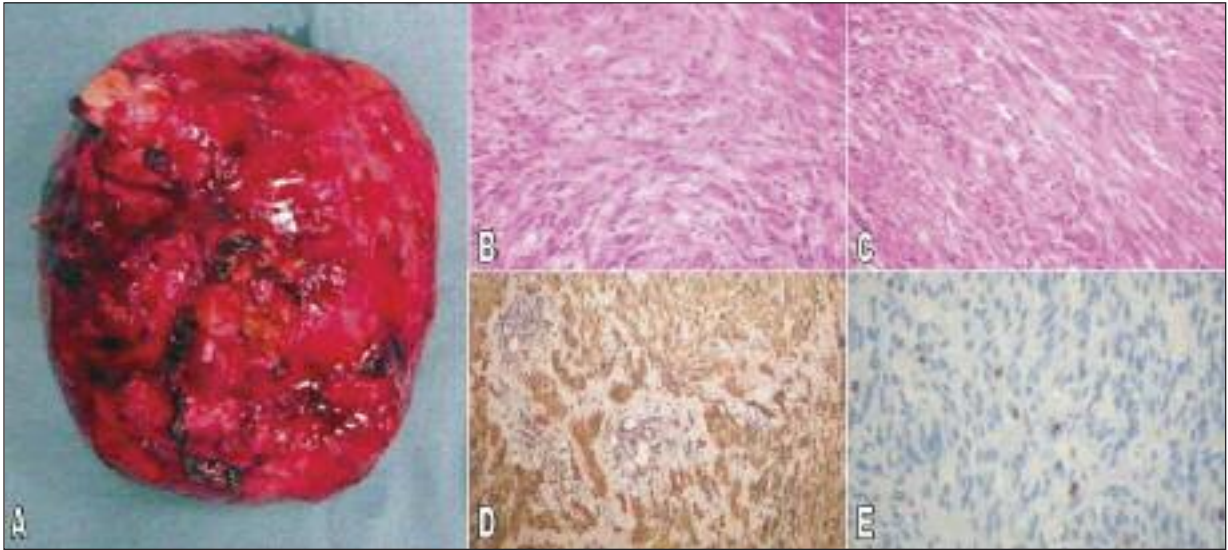


FIGURE 2: (A): Macroscopic view of the tumor, (B): Normocrom hypercellular area with spindle-shaped tumor cells, (HE, x200). (C): Hypocellular area with spindle-shaped tumor cells, (HE, x200). (D): The tumor cells were immunohistochemically positive for S-100 protein, (IHC, x100). (E): 2-3% of tumor cells were positive for Ki-67 (MIB-1), (IHC, x100).

areas of spindle-shaped cells, densely populated, forming a fascicular, storiform and whorled-growth pattern. Foci of nuclear palisading were present with no evidence of Verocay bodies. Immunohistochemically, the tumor cells were strongly and diffusely positive for vimentin and S-100 protein. Proliferating cell-associated antigens Ki-67 (MIB-1) index was 2-3% in all tumors (Figure 2).

Post-operative course was uneventful. She promptly regained normal respiration function. Operation also resulted in satisfactory pain relief with good healing of the surgical wounds. The patient was discharged from our clinic after 7 days. At present, in postoperative month one, the patient continues to display no evidence of respiratory distress and satisfactory pain relief.

DISCUSSION

Schwannoma, a relatively rare retroperitoneal tumor, has a reported incidence of only 0.5-1.2%.⁴⁻⁶ The low frequency of this tumor and the lack of specific instrumental signs and objective symptoms (since it develops in a deep and broad region, the retroperitoneum) make presurgical diagnosis extremely difficult.^{7,8} Clinically, early detection of the

tumor is difficult unless it enlarges and becomes palpable or compresses the surrounding organs. To our knowledge, no instance of schwannoma in this region and pulmonary atelectasis due to the elevation of the diaphragm was reported in the literature.

The pathologist who is aware of this clinical picture and faced with a tumor of probable nerve origin that possesses atypical histologic features, such as increased cellularity, hyperchromatism, nuclear pleomorphism, and mitotic activity, should be aware of a common pitfall-misdiagnosing a CS as a malignant MPNST. MPNST, is usually more cellular, associated usually with a higher degree of anaplasia, lacks the thick walled hyalinised blood vessels and usually demonstrates only focal S-100 protein positivity on immunohistochemistry.⁹ Although S-100 protein can be identified in 50-90% of MPNSTs, staining is typically focal and limited to a small number of cells. Strong, diffuse immunoreactivity for S-100 protein always suggests benign schwannoma, including a cellular variant other than MPNST. Ki-67 protein is expressed in the proliferative phase (G1, S, G2) of the cell cycle. In MPNST, Ki-67 protein indexes were reported in 5-65%; in con-

trast, indexes of CS were typically less than 10%.^{2,10} These immunohistochemical analyses are beneficial for the differentiation between CS and MPNST.

Differentiation of CS from malignant tumors is possible if strict criteria are used on histological and immunohistological analyses, thus avoiding misdiagnosis. Misinterpretation of a cellular schwannoma

may cause the patient suffer from side effects of unnecessary overtreatment with radiation and chemotherapy.

In addition, long-term follow-up is certainly needed for prognosis because mitotic count significantly correlates with the incidence of tumor recurrence and thus close follow-up is strongly suggested.^{2,11-13}

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