Intrathoracic Schwannoma Presented with Hemothorax

Schwannom ve Hemotoraks / Schwannoma and Hemothorax

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Özet
İntratorasik schwannomalar nöral kolifta bulunan Schwann hücrelerinden köken alan benign ve asemptomatik tümörlerdir. Intratorasik schwannomaların spontan hemoraji sonucu hemotoraks yol açması nadir görülen bulgudur. Nefes darlığı ve sağ kolda uyuşma şikayetleri bulunan 17 yaşındaki bayan hastaya çekilen ön-arka akciğer grafisinde sağ hemitoraksta masif pleval efüzyon saptanmış olup yapılan torasentezde hemotoraks ile uyumlu sıvı alınması üzerine tüp torakostomi uygulandı. Çekilen kontrol ön-arka akciğer grafisinde sağ hemotoraks kitle tespi edildi, bilgisayarlı toraks tomografisinde interkostal sinirlerden köken alan 76 x 104 mm boyutlarında kitle tespi edildi. Rezeke edilen kitenin histopatolojik inceleme sinde schwannoma olduğu raporlanan hasta reseksiyon sonrası on dört aydır hasta tılsız hayata devam etmektedir.

Anahtar Kelimeler
İntratorasik Schwannom; Hemotoraks; Torakotomi; İnterkostal

Abstract
Intrathoracic schwannomas are typically benign and asymptomatic tumors that originate from the Schwann cells of a neural sheath. Hemorrhage from intrathoracic schwannomas is an uncommon finding. We present the case of a 17 year-old girl who had dyspnea and numbness of the right arm. Chest x-ray showed a right-sided massive pleural effusion and exploratory puncture showed hemothorax. After tube thoracostomy and drainage of the bloody effusion, a mass was observed in the right upper hemithorax. Chest tomography revealed a 76x104 mm mass arising from the fourth intercostal nerve. The tumor was successfully resected and, on histopathologic examination it was reported as a schwannoma. After resection, the patient has remained problem-free for fourteen months.

Keywords
Intrathoracic Schwannoma; Hemothorax; Thoracotomy; Intercostal
Introduction

Neurogenic tumors are commonly found, especially in the posterior mediastinum or in the chest wall, and have a variety of clinical features [1]. Intrathoracic schwannomas are typically benign and asymptomatic tumors that originate from the Schwann cells of a neural sheath [1,2]. Hemorrhage from intrathoracic neurogenic tumors is extremely rare [3]. A review of the English language medical literature found only 7 previous case reports describing hemothorax due to intrathoracic schwannoma.

Case Report

A 17 year-old girl was admitted to our hospital with dyspnea and numbness of the right arm. She had these complaints for a period of fifteen days. On physical examination there was no breathing sounds in the right hemithorax; there was no neurological malfunction except numbness of the right arm. Chest radiography revealed a massive, right-sided pleural effusion and thoracentesis was performed. Exploratory puncture of the right thoracic cavity revealed a bloody pleural effusion and right tube thoracostomy was performed on the patient due to hemothorax. Following the drainage of 750 cc hemothorax, a second chest x-ray was performed, which showed a smooth, rounded mass on the upper-right part of the hemithorax (Figure 1a). We planned a thoracic computerized tomography (CT), which revealed a regular, round mass (86x73 mm) arising from the right thoracic wall, which appeared to displace the trachea to the left side of the mediastinum (Figure 1b). Those details of the tumor were obscured by the pleural effusion. A decision of transthoracic fine-needle aspiration biopsy was made and pathologic examination of specimens indicated a mesenchymal tumor. Hence, we performed chest magnetic resonance imaging (MRI) to clarify the location and characteristics of the tumor. On examination of the chest MRI a huge tumor mass (74x76x104 mm) with high signal intensities was determined on T2-weighted images. On positron emission tomography (PET) examination, the mass had a maximal standard uptake value of 7.26.

Discussion

Both somatic and autonomic nervous systems are found throughout the thorax and concentrated in the paravertebral sulcus region, known as the posterior mediastinum [1]. Most of the peripheral nerve tumors in the thorax are located in the posterior mediastinum, and 12% to 21% of all mediastinal tumors are neurogenic [1]. Intrathoracic schwannomas are uncommon tumors that originate from the Schwann cells of a neural sheath.

In general, schwannomas are slow-growing and asymptomatic tumors except for the compression of neighboring structures [4]. They rarely develop into malignant tumors and the risk of malignancy in a nerve sheath tumor is very small (2% - 5%) [1,2]. On analyzing a series of neurogenic tumors of the chest, Yamaguchi et al. [1] reported that only one (1.7%) of 60 patients had malignant schwannoma. If the patient has a history of radiation exposure or Von Recklinghausen's disease, the malignancy risk increases to 10% - 20% [2]. Malignancy is suggested by features such as mitotic activity, necrosis, nuclear pleomorphism and invasion of surrounding tissues and vascular structures [2]. In our case, there were no malignant features and the diagnosis of schwannoma was made without difficulty.

The majority of intrathoracic neurogenic tumors are asymptomatic [1,4]. However, as they become larger in size, these space-occupying lesions can produce local compression symptoms in adjacent structures such as bone erosion and spinal cord involvement [1]. Schwannomas rarely manifest with hemothorax. White et al. [5] reviewed 57 schwannoma cases, of which only 3 were in the chest wall and none presented with hemothorax. In a review the English language medical literature, there were only 7 cases of intrathoracic schwannoma appeared with hemothorax [2,3]. Both Morimoto et al. [2] and Lee et al. [3] described intrathoracic schwannoma cases presented with hemothorax and treated successfully by surgical excision. The causes of bloody pleural effusion were reasoned as bleeding caused by external trauma or a weak locus within the tumor [2]. In our patient, there was no history of trauma; thus, we suspected that some weak locus within the tumor might have been ruptured and caused hemothorax and shortness of breathing.

Typically, schwannomas show equal or lower signal intensities than muscles on T1-weighted MR images. Moreover, they show inhomogeneous high signal intensities on T2-weighted MR images [2]. Morimoto et al. [2] reported a similar case of schwannoma presented with hemothorax and they emphasized the importance of thoracic MRI in preoperative evaluation. Our case also had similar MRI findings to those described in the literature.
Thoracotomy was necessary not only as a diagnostic procedure, but also to ensure completeness of excision and to free a trapped lung [3]. In an analysis of 60 cases, open surgery, including thoracotomy and sternotomy, was performed in 51 patients (85 %), whereas only 9 patients (15 %) were treated using video-assisted thoracic surgery [1]. It was emphasized that best approach would be standard posterior thoracotomy, located one or two intercostal spaces above or below the tumor [1]. In our case, our approach was also standard posterior thoracotomy, one costal space below the mass.

Intraspinal extension of neurogenic tumors requires a combined posterior spinal and thoracic approach for safe resection [6]. A single-stage procedure is recommended because thoracic manipulation of the tumor can produce bleeding within the tumor; hemorrhagic expansion of the tumor within the fixed space of the spinal canal can result in cord compression and paralysis [6]. We performed our case with the help of neurosurgeons although the tumor was not extended to the intraspinal part of the spinal cord. Complete excision of benign schwannomas is considered curative and reported recurrence is uncommon [4].

**Conclusion**

In conclusion, we report a rare case of intrathoracic schwannoma, originating from intercostal nerve, presenting with hemothorax. Immediate drainage and complete surgical excision of the tumor successfully restored normal pulmonary function. In hemothorax patients a possible underlying neoplasm should always be considered. Consequently, family physicians, neurologists, and thoracic surgeons should be aware that spontaneous massive hemothorax in a patient may result from neurogenic tumors such as schwannoma.

**References**