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Schwannoma arising from the right phrenic nerve

Sağ frenik sinir kökenli schwannoma

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Schwannoma of the intrathoracic phrenic nerve is a rare presentation. Routine chest X-ray examination of a 43-yearold asymptomatic man revealed a smooth, round, abnormal shadow in the right hilum adjacent to the heart. Computed tomography showed a regular mass in the right cardiophrenic junction, measuring 7x5x5 cm. On exploratory right thoracotomy, there was a firm mass involving the right phrenic nerve at the level of the cardiophrenic junction. The mass was peeled off along the phrenic nerve from the pericardium. Histopathology showed a benign schwannoma originating from the right phrenic nerve. Temporary mild right diaphragmatic eventration occurred after the operation but there was no respiratory distress. The patient's respiratory function returned to normal nine months after the operation.

Key words: Neurilemmoma/surgery; peripheral nervous system neoplasms; phrenic nerve.

Neurogenic tumors represent one of the common causes of mediastinal masses. In adults, the majority of these are benign and found incidentally. They can originate in any neurogenic structure in the mediastinum, including the sympathetic or parasympathetic chain, intercostal nerves, and spinal ganglia. A schwannoma is a tumor of the nerve sheath. These tumors develop from Schwann cells. It can be malignant or benign. Management of these tumors is primarily surgical excision, which is curative with recurrences being rare.^[1]

CASE REPORT

A 43-year-old male patient without any symptoms was referred to our clinic after a routine evaluation. Chest radiography showed a smooth, round, abnormal shadow in the right cardiophrenic corner adjacent to the heart, suggesting a pericardial cyst, hydatid cyst, any cystic lesion or tumor (Fig 1a). Computed tomography showed a regular mass in the right cardiophrenic junction, measuring 7x5x5 cm. On exploratory right thoracotomy, there was a firm, well-circumscribed, completely encapsulated mass involving the right phrenic nerve

İntratorasik frenik sinir kökenli schwannomalar çok nadirdir. Kırk üç yaşındaki asemptomatik erkek hastanın rutin akciğer grafisinde, kalp komşuluğunda, sağ hilusta, yuvarlak gölge veren bir lezyon saptandı. Bilgisayarlı tomografide, sağ kardiyofrenik bileşkede, 7x5x5 cm boyutlarında düzenli bir kitle görüldü. Sağ torakotomi ile yapılan eksplorasyonda, kardiyofrenik bileşke düzeyinde sağ frenik siniri tutan sert bir kitleye rastlandı. Tümoral kitle sağ frenik sinirden sıyrılarak çıkarıldı. Histopatolojik tanı sağ frenik sinir kaynaklı benign schwannoma olarak bildirildi. Ameliyat sonrası dönemde solunum bozukluğuna neden olmayan, orta dereceli diyafragma evantrasyonu gözlendi. Hastanın solunum fonksiyonları ameliyattan dokuz ay sonra normale döndü.

Anahtar sözcükler: Nörilemoma/cerrahi; periferik sinir sistemi neoplazileri; frenik sinir.

at the level of the cardiophrenic junction (Fig 1b). The mass exhibited an extraparenchymal and extrapericardial location. There were no other parenchymal lesions or mediastinal adenopathy. The pericardium appeared not to be involved. The mass was peeled off along the phrenic nerve from the pericardium. Histopathology showed a benign schwannoma originating from the right phrenic nerve. Right diaphragmatic eventration occurred after the operation but there was no respiratory distress. The postoperative course was uneventful. The patient was discharged on the fifth postoperative day and his respiratory function returned to normal nine months after the operation.

DISCUSSION

The most common masses occurring in the posterior mediastinum are neurogenic tumors. Among these, neurofibromas and schwannomas originating from the peripheral nerve sheath are the most common. Neurogenic tumors represent 20% of all adult and 35% of all pediatric mediastinal neoplasms. The majority of these lesions in adults are benign, asymptomatic, and

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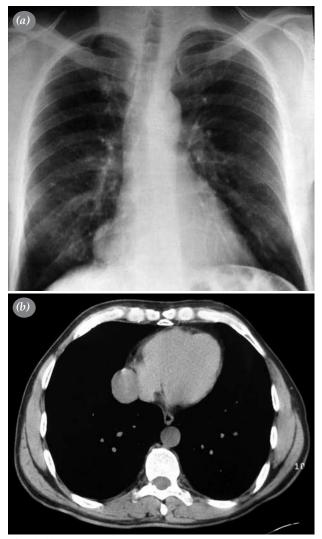


Fig. 1. (a) Chest radiography shows a smooth, round, abnormal shadow in the right cardiophrenic corner adjacent to the heart. **(b)** Computed tomography scan shows a regular mass in the right cardiophrenic junction, measuring 7x5x5 cm.

incidentally found.^[2] Schwannomas are benign, slowgrowing neurogenic tumors that arise from the sheath of a spinal nerve root or any thoracic nerve and may cause extrinsic compression to the associated nerve. They can arise anywhere in the body. These tumors develop from Schwann cells. Malignant transformation is very uncommon. Intrathoracic schwannomas usually arise from the intercostal and sympathetic nerves and phrenic nerve origin is very rare. In a review of 138 mediastinal tumors, only one originated from the phrenic nerve.^[3] There are several case reports of schwannoma arising from the phrenic nerve.^[3-5] In our case, schwannoma originated from the right phrenic nerve and the diagnosis was made by histopathologic examination. None of the reported cases were correctly diagnosed on the basis of imaging studies.

Benign schwannomas are generally asymptomatic. If the tumor invades the nerve block, symptoms occur due to nerve dysfunction. Site pain occurs with intercostal nerve block, diaphragmatic eventration occurs with phrenic nerve block. Schwannoma arising from the phrenic nerve can cause unilateral diaphragmatic paralysis. In a review of 142 patients with unexplained diaphragmatic eventration, phrenic nerve involvement by the tumor was diagnosed in only five patients. Of these, four patients had bronchogenic carcinoma and one patient had metastatic hepatocellular carcinoma.[6] Surgery is indicated in symptomatic eventration or in cases of very large eventrations, even if asymptomatic. Plication of the diaphragm is the procedure of choice. Reinforcement with a synthetic mesh may be required. It is suggested that schwannoma arising from the phrenic nerve be removed completely and that cutting one side of the phrenic nerve would be easy if the patient has normal respiratory function.^[5] If schwannoma is benign, it is not necessary to cut the phrenic nerve. There was no diaphragmatic eventration in our case. We peeled off the tumor from the phrenic nerve and avoided cutting. After the operation, temporary diaphragmatic eventration occurred.

In conclusion, schwannoma arising from the intrathoracic phrenic nerve is rare, and to our knowledge, only a few cases have been reported in the literature. Its management is primarily surgical excision.

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