Hemangiopericytoma located in the mediastinum: a case report

Mediasten yerleşimli hemanjiyoperisitoma: Olgu sunumu

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Hemangiopericytomas, which arise from cells (pericytes) that are wrapped around the pericapillary arterioles, are commonly located in the soft tissue of the thigh and retroperitoneum. Hemangiopericytoma in the mediastinum is extremely rare. Fewer than 20 cases were reported and while one fourth of them was benign and the remainder was malignant. The malignant potential of hemangiopericytoma is a distinctive characteristic when compared to the other vascular tumors. A high-signal area on T2-weighted magnetic resonance imaging and a massive bleeding mass during the operation are the characteristic features of hemangiopericytoma. Complete resection is the only effective treatment option for this vascular slow-growing tumor. In this article, we report a case of a 23-year-old male who was treated by complete surgical resection of the tumor. Details of clinic, pathologic and radiographic features are presented.

Key words: Hemangiopericytoma; mediastinum; vascular tumor.

Hemanjiyoperisitomalar perikapiller arteriollerin etrafını saran hücrelerden (perikist) köken alan, genellikle retroperiton ve uyluğa ait yumuşak dokuda yerleşen tümörlerdir. Mediastinal yerleşimli hemanjiyoperisitoma oldukça enderdir. Bildirilmiş 20'den az olgu bulunmaktadır ve bunların dörtte biri benign iken diğerleri maligndir. Hemanjiyoperisitomanın malign olma potansiyeli diğer vasküler tümörler ile karşılaştırıldığında ayırt edici bir özelliktir. T2 ağırlıklı manyetik rezonans görüntülemede yüksek sinyalli bir alan ve ameliyat sırasında masif kanamalı bir kitle hemanjiyoperisitomanın karakteristik özelliklerindendir. Bu yavaş büyüyen vasküler tümörün tek etkili tedavi seçeneği tam rezeksiyondur. Bu yazıda tam cerrahi rezeksiyon uygulanarak tedavi edilen 23 yaşında erkek bir olgu klinik, patolojik ve radyografik özellikleri ile sunuldu.

Anahtar sözcükler: Hemanjiyoperisitoma; mediasten; vasküler tümör.

Hemangiopericytoma is an uncommon and potentially malignant mesenchymal tumor that occurs most commonly in the skin, the subcutaneous soft tissues, muscles of the extremities or retroperitoneum, but rarely in the lung, trachea or the mediastinum.

The tumor apparently arises from spindle-shaped pericytes that lie at the basement membrane of capillary blood vessels. Although half of the patients are asymptomatic, the others' signs and symptoms are protean, such as dysphagia, Pancoast syndrome like features and occurrence of a spontaneous hemothorax.

Herein, a completely resected case of primary mediastinal hemangiopericytoma is presented with its clinic, pathologic and radiographic features.

CASE REPORT

A 23-year-old man was referred to our institution with an abnormal appearance in his chest roentgenogram. He had no symptoms related to the mass and his physical examination was normal. Laboratory studies were essentially within the normal limits. A mass shadow with a well-defined margin in the left hilum on the posteroanterior view was shown on the X-ray. A computed tomography (CT) scan of the chest revealed a mass measuring 4x4x3 cm, located in the paratracheal area and proximally of the left main pulmonary artery. The mass showed the same enhancement with the aorta and the pulmonary artery and the border with the left main pulmonary artery was not well-defined. To rule out any pulmonary arterial aneurisms, a pulmonary angiography was performed. The result indicated that the lesion had no pulmonary arterial association.

The magnetic resonance imaging (MRI) showed a high-intensity mass not invading the aorta in the T1 and T2-weighted images (WIs) (Fig. 1). There was no evidence suggesting another primary tumor or metastasis, and a surgical management was suggested.

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The patient underwent a left-sided thoracotomy. During the thoracotomy, a 4-cm solid mass was found originating from the mediastinum, proximally of the left main pulmonary artery, but without a bronchial attachment or any significant arterial supply. There was an involvement of the surrounding lung parenchyma. Despite the persistent bleeding during the dissection, the tumor was resected. The frozen section of the lesion suggested a thymoma, a benign spindle cell tumor or another low grade soft tissue sarcoma; and therefore, there was no need for any extended resection.

The tumor mainly consisted of fleshy fragments. They were not circumscribed and did not show any extensions into the lung. Histologically; there was a cellular mesenchymal tumor composed of round or fusiform cells demonstrating focal cellular pleomorphism. It was rich in thin-walled branching vessels forming a staghorn vascular pattern. There was no necrosis. The mitotic activity was approximately three per 10-high-power microscopic fields and the Ki-67 immunoreactivity was around 5%. The tumor was reported to be a "hemangiopericytoma having low malignant potential" (Fig. 2).

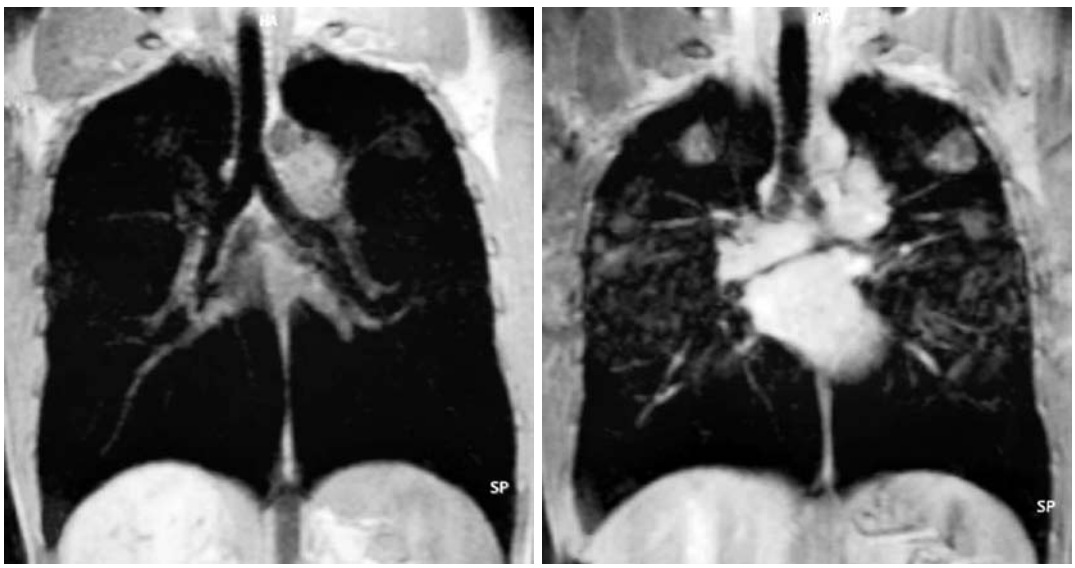
The postoperative course was uneventful and patient was discharged on the sixth postoperative day. Two weeks after the operation, a positron emission tomography (PET) scan was performed to rule out the possibility of an incomplete resection and distant metastases. There was no evidence of any of these pathologies. A postoperative chemo-radiotherapy was not recommended. Today, 14 months after the operation, the patient is well and there is no evidence of any recurrence or metastasis.

DISCUSSION

Hemangiopericytoma is an uncommon, mesenchymal and potentially malignant tumor arising from the capillary pericytes. In our case, it was in the mediastinum, which is an extremely rare location for this kind of a tumor.

Hemangiopericytoma was initially described by Stout and Murray.[1] The skin, musculoskeletal system and retroperitoneal areas are most commonly affected. Only a few cases of tumor originating from the mediastinum have previously been reported. The diagnosis of extra-pulmonary tumor is difficult. Most of the patients are asymptomatic. A non-calcified solitary mass with a smooth margin is discovered on the chest-X-ray. Because this is a vascular tumor usually demonstrating hypervascularity, MRI or angiography are more effective diagnostic tools rather than a CT scan. In our case, the angiography was normal and the MRI demonstrated a high intensity on T_1 and T_2-WIs. Kusumoto et al.[2] have reported that the MRI shows a characteristic high signals on T_2-WIs and this feature can be useful to detect an early recurrence or metastatic lesions.

The most important fields of use of the PET include evaluating solitary pulmonary nodules and the mediastinal staging of lung cancer. Its mechanism depends on the increased usage of glucose for the increased mitotic activity in cells like the cancer cells. Although there is no information about using PET for hemangiopericytoma in the literature, we chose to use it in our case because the mitotic activity rate of the lesion was relatively higher than the other benign lesions.



Şekil 1. Contrast-enhanced T_1 and T_2 weighted magnetic resonance demonstrating a hyperintense mediastinal mass.

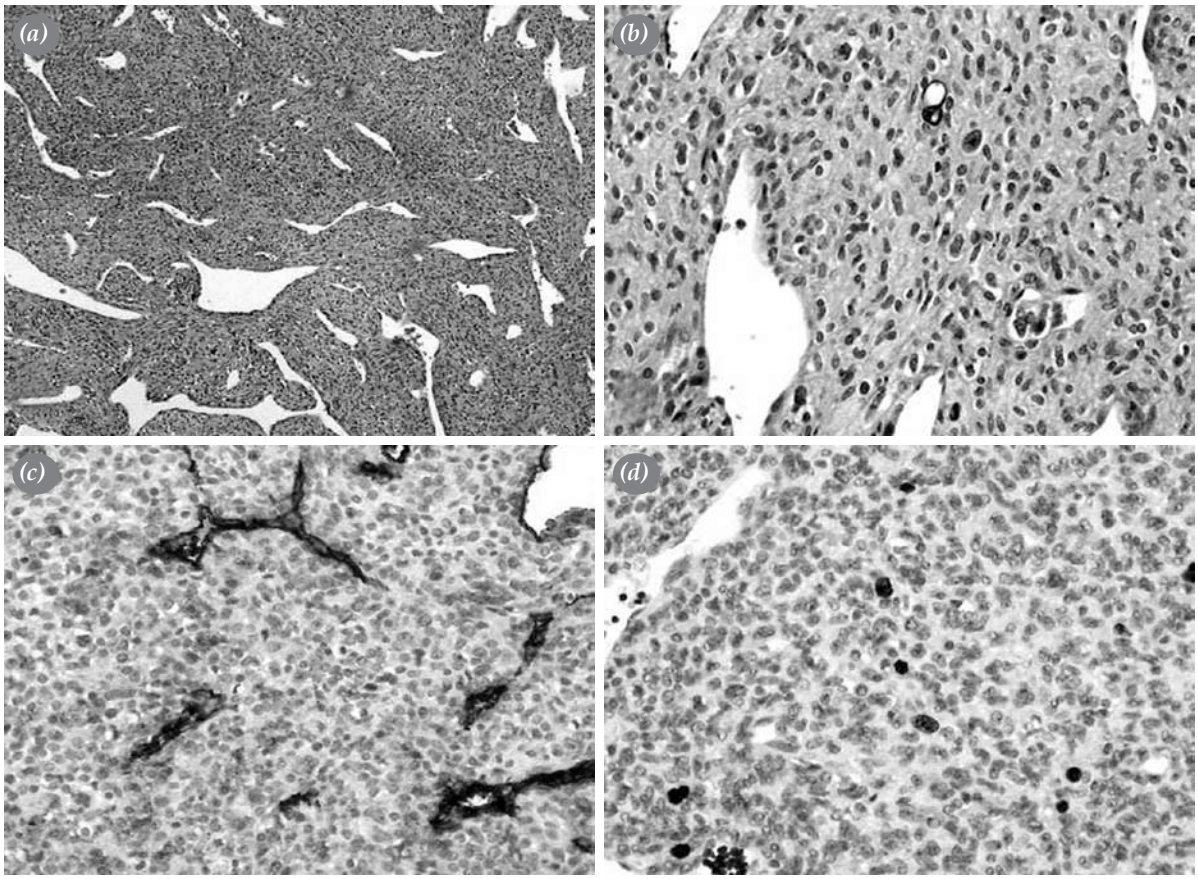
The histological differential diagnosis of hemangiopericytoma includes many mesenchymal tumors, such as the solitary fibrous tumor and the synovial sarcoma.[3] The diagnosis can be difficult in the preoperative stage. A frozen section may not be reliable.[4] In our case, the frozen section of the lesion demonstrated a thymoma, benign spindle cell tumor or another low-grade soft tissue sarcoma.

The histopathological examination plays a very important role in the diagnosis of the tumors with four or more mitoses per 10 high-power microscopic fields, prominent cellular pleomorphism, increased cellularity and areas of hemorrhage and necrosis are usually malignant. Immunohistochemically, hemangiopericytomas are known to be stained for vimentin type 4 collagen and they were not stained for type 8-related antigen, S-100 protein, neuron specific enolase, carcinoembryonic antigen, desmins, laminin and cytokeratins.

Surgical resection is the first choice of treatment.[3] Hansen et al.[4] stated that it was necessary to consider all hemangiopericytomas as malignant and perform extended surgery, however the criteria for determining the area of resection has not been established yet. Chemotherapy, radiotherapy or both have been recommended, however, it is generally considered to be almost ineffective.[3] On the other hand, combination therapy or single therapy with Adriamycin was reported to be effective against metastases.[5] In order to allow a safe and complete removal of the lesion; Jalal and Jeyasingham[6] have suggested that preoperative radiotherapy to large hemangiopericytomas located on the chest wall, significantly reduced their vascularity. Morandi et al.[7] have suggested preoperative percutaneous embolization of hypervascular mediastinal tumors.

The five-year survival rate of patients with hemangiopericytomas originating in any organ has been reported to be approximately 85%, whereas the survival rate of patients with a tumor of pulmonary or mediastinal origin is comparatively low.[4]

In conclusion, hemangiopericytoma, a vascular, slow-growing tumor, is rarely encountered in the mediastinum. A radical surgical excision is necessary



Şekil 2. (a) Histologically, hemangiopericytoma is rich from thin walled vessels and exhibits staghorn vascular pattern (H-E x 50). **(b)** Tumor is cellular and contains scattered pleomorphic cells (H-E x 200). **(c)** CD 34 immunostaining highlight the vascularity (Avidin-biotin immunostaining x 100). **(d)** Ki-67 immunoreactivity is about 5% (Avidin-biotin immunostaining x 200).

for the treatment, because other treatment modalities are ineffective. Based on our experiences from this case, we want to emphasize that the characteristic high signal on T2-WIs in the preoperative MRI and a massive intraoperative bleeding. Local or distant recurrence is commonly seen and a long-term follow-up is required.

KAYNAKLAR

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