Unexpected Etiology in Pericardial Effusion: Malignant Fibrous Histiocytoma: Case Report

Perikardiyal Efüzyonda Beklenmedik Etiyoloji: Malign Fibröz Histiyositoma

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ABSTRACT Although malignant fibrous histiocytoma is the most widely seen malignant soft tissue tumor in adults, mediastinal localizations are uncommon. In this report, we aimed to present a patient admitted to hospital with complaints of hypotension, vertigo, arrhythmia and operated under emergency conditions for severe paericardial effusion leading to right atrial collapse and diagnosed as malignant fibrous histiocytoma.

Key Words: Histiocytoma, malignant fibrous; mediastinum; soft tissue neoplasms; heart neoplasms

ÖZET Malign fibröz histiyositoma erişkinlerde malign yumuşak doku tümörleri arasında en sık görülen tümör olmasına rağmen, mediastinal yerleşimi nadirdir. Bu yazıda, hipotansiyon, vertigo, yorgunluk ve aritmi yakınmaları ve sağ atrial kollapsa neden olan ve ileri derecede perikardiyal efüzyon nedeni ile acil perikardiyal tüp drenajı uygulaması sırasında tanı konulan bir malign fibröz histiyositoma olgusu sunulmaktadır.

Anahtar Kelimeler: Malign fibröz histiyositoma; mediasten; yumuşak doku tümörleri; kalp tümörleri

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The most common soft tissue sarcoma is malignant fibrous histiocytoma. They may originate from the thoracic wall, lungs, mediastinum and other soft tissues.[1-5] They are most commonly seen in thoracic wall, rarely in the mediastinum. Surgery and radiotherapy and chemotherapy following surgery may yield good results in these tumors.[4] Because sarcomas have better prognosis than carcinoma, 5-year survival rates vary between 40-70%.[1-3] Despite tissue and organ metastases are seldom, better results can be achieved as a result of surgical treatment of existing invasions[1,4,6]

Here, we aim to present a case misdiagnosed and underwent tube drainage due to pericardial effusion and then diagnosed as malignant fibrous histiocytoma localized in the right ventricle.

CASE REPORT

In the examination of a 67-year-old male patient who admitted to cardiology clinic with complaints of hypotension, vertigo and an ever-increasing state of fatigue and arrhythmia. Arterial pressure was 100/60 mmHg, rhythm was atrial fibrillation with rapid ventricle response in electrocardiography (rate 160/min), and advanced pericardial effusion leading to right atrial collapse in echocardiography were seen in physical evaluation and monitoring.

Emergency pericardial tube drainage was performed through the subxyphoid approach under local anesthesia. Approximately 600 ml serohemorrhagic and fibrinous fluid was drained. A mass-like hard tissue, adjacent to the inferior margin of the heart and the pericardium, was found by digital examination made from the pericardial window. Pericardial fluid samples for cytologic, microbiologic and biochemistrical examination and biopsy sample for pathological examination were collected. No specific findings were detected upon examination of these samples. Patient was entubated and the mediastinum was explored by median sternotomy under general anesthesia. We detected a mass about 8x10 cm in size, on the right ventricle wall facing the diaphragm (Figure 1). Because it was not possible to surgically remove the mass, the operation was ended after taking a frozen tissue sample for pathological examination. The frozen sample was reported as malignancy. Silicone drains were placed in the mediastinum, sternum incision was closed with standart techniques. Patient was in sinus rhythm and arterial pressure was 150/80 mmHg postoperatively. Histopathologic examination revealed that the tumor consisted of malignant cells with marked atypia, multinuclear, markedly pleomorphic cells. The biopsy result was reported as malignant fibrous histiocytoma. Patient was consultated by oncologists and they decided radiotherapy. No marked decrease was observed in tumor sizes after applying 25-days of radiotherapy, however a clinical improvement was observed in the patient's symptoms. The patient is still being followed up with stable clinical findings at the sixth month postoperatively.

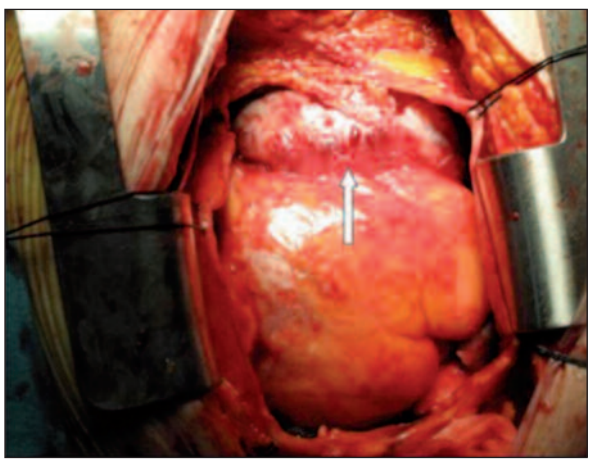

FIGURE 1: Intraoperative image of malignant fibrous histiocytoma.
(See color figure at http://cardivascular.turkiyeklinikleri.com/)

DISCUSSION

Malignant fibrous histiocytoma is the most commonly encountered soft tissue sarcoma in adults and it makes metastases to the lungs, bones and liver respectively. In their study, Belal et al. reported that malignant fibrous histiocytoma 47% originated from the lower extremities, 18% from upper extremities, 16% from the head and neck region, 9% from the body, 5% from the pelvis and 5% from other regions.[7] Many tumors or cysts exhibiting various histopathological features might arise from the anatomic structures in the mediastinum. Masses in the mediastinum reveal symptoms after they cover a space and grow to compress the vital structures. Despite the fact that malignant fibrous histiocytoma is one of the mostly-encountered tumors of the soft tissue, they are rarely seen in the thorax and primarily in the mediastinum.[1,3-6]

It has been reported that cardiac sarcomas occur more frequently in the right side of the heart, especially in the right atrium. However, cardiac malignant fibrous histiocytoma is most frequently located in the left atrium, but we observed the tumor in the right ventricle which is a very rare localization.[8,9] Twenty-nine percent of patients have pericardial effusions at presentation.[10] Our patient had a severe pericardial effusion leading to right atrial collapse.

In malignant fibrous histiocytoma, wide excision is the primary method of treatment due to high local recurrence and remote metastasis rate. Because they exhibit a biological behavior different than the carcinoma, they can be surgically removed even if they cause mediastinal invasion. Even in some malignant cases that cannot be fully resected, surgery has benefits like the removal of the compression on mediastinal structures and ensuring the collection of sufficient amount of material for histological diagnosis. In our case, because the mass exhibited an advanced degree of invasion to the right ventricle on the cardiac base, surgical excision was considered unfavourable. Postoperative radiotherapy was performed.

In conclusion, although severe pericardial effusion is a rare presentation for mediastinal tumors, advanced examination and treatment options should always be considered for malignant tumors, in cases that cannot be diagnosed as a result of pericardial fluid analyses. Radiotherapy practice may cause symptomatic improvement in cardiac malignant fibrous histiocytoma that can't be diagnosed and surgically treated at an early stage due to rapid tumor growth.

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