Paracardiac Giant Cystic Desmoid Tumour: Case Report

Parakardiak Dev Kistik Desmoid Tümör

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ABSTRACT Desmoid tumours, also called aggressive fibromatosis, are an extremely rare type of tumour arising from musculoaponeurotic tissues. Their etiology is still unknown. They are usually benign, but local invasion and relapsing are frequent. Here we report the case of a 45-year-old male patient with widespread invasion of a desmoid tumour involving superior-posterior mediastinum, retroperitoneum and right lumbocostal region. The tumour was inoperable because of important vascular and cardiac invasions. Despite the intra-abdominal location of most desmoid tumours, involvement of the superior mediastinum and their differential diagnosis among the retroperitoneal-mediastinal masses should be definitely kept in mind. Transthoracic ecocardiography should be considered as a first method for the assessment of mediastinal invasion.

Key Words: Fibromatosis, aggressive; echocardiography

ÖZET Agresif fibromatozis olarak da adlandırılan desmoid tümörler, musküloaponevrotik dokulardan kaynaklanan çok nadir görülen bir tümör tipidir. Etiyolojisi halen bilinmemektedir. Genellikle iyi huylu olup, lokal invazyon ve nüks sıktır. Bu yazıda superior-posterior mediasteni, retroperitoneal ve sağ lumbokostal bölgeyi içeren yaygın invazyonun görüldüğü desmoid tümörlü 45 yaşındaki erkek hastayı sunuyoruz. Önemli vasküler ve kardiyak invazyon nedeniyle tümör inoperabl kabul edildi. Desmoid tümörlerin çoğu intra-abdominal yerleşime sahip olmasına rağmen, süperior mediasten tutulumu ve retroperitoneal-mediastinal kitleler arasındaki ayırıcı tanıları mutlaka akılda bulundurulmalıdır. Transtorasik ekokardiyografi mediasten invazyonu değerlendirmesinde ilk metot olarak düşünülmelidir.

Anahtar Kelimeler: Fibromatozis, saldırgan; ekokardiyografi

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Desmoid tumours are rare neoplasms originating from connective tissue. Despite their benign histopathology, these inoperable tumours display malignant behavior in advanced stages as they invade vital organs and veins. Here, we present the case of a 45-year-old male patient who had previously been operated several times due to retroperitonel desmoid tumour, and who over the years had slow but widespread invasion of the recurrent desmoid tumour involving as wide a region as to the superior mediastinum and the right lumbocostal skin region.

CASE REPORT

A 45-year-old male was referred to our emergency department due to severe typical angina, dyspnea and palpitation. On physical examination, there was no pathologic finding except a huge palpable mass at the right lateral lumbocostal region lasting for six months. In the patient history, it was discovered that he underwent surgical operations due to retroperitoneal desmoid tumour in 1987 and 1991. His electrocardiography was compatible with supraventricular tachycardia with heart rate 200 beat/min. A chest X-ray showed cardiac enlargement, cardiac shift to the left side, right pleural effusion, right diaphragmatic elevation and mass contour under the diaphragm (Figure 1). The patient's transthoracic echocardiograpy (TTE) indicated that the dimension of the left ventricle and its systolic function were normal. Huge cystic mass, 10x15 cm in size, compressing the left atrium was detected at the back of the heart (Figure 2). In order to see whether the mass involved other regions, thoraco-abdominal computed tomography was performed. Heterogeneous lobulated cystic mass extending from the superior mediastinum to upper retroperitoneum and the right lateral lumbocostal skin region was found. The maximum size of mass was 15x22x23 cm in the thorax. Esophagus, descending aorta, trachea and main bronchial structures were surrounded and right kidney was displaced to inferior by the mass (Figure 3). The coronary angiography taken to exclude coronary arterial disease was normal. The patient's supraventricular tachycardia was successfully treated with the radiofrequency ablation method. No tachycardia attack was observed in his 24-hour Holter monitor follow-up. It was believed that the patient had desmoid tumour recurrence, which was confirmed by transcutaneous mass biopsy. Histopathologically, spindle cells consisting of dense collagen fibers and fibroblasts were observed (Figure 4). The patient was thought as inoperable because of vital organ invasions. Adjuvant radiation therapy was planned. No meaningful change in tumour size was detected in the patient's 3-month follow-up.

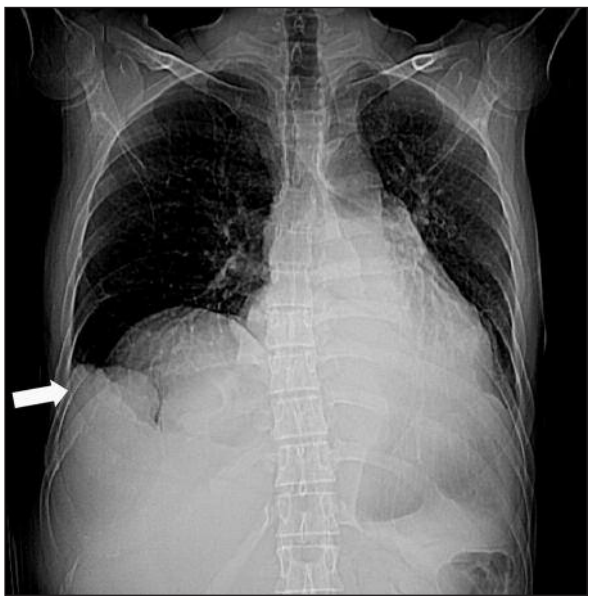

FIGURE 1: Chest X-ray shows right pleural effusion and remarkable mass contour under diaphragm (arrow).

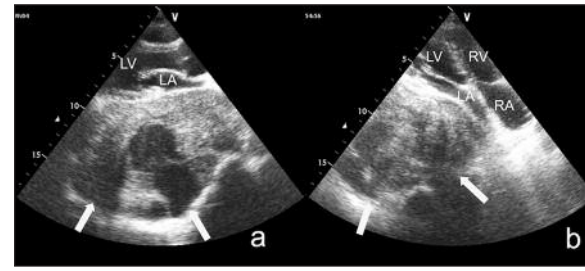

FIGURE 2: Transthoracic Echocardiography indicates huge cystic mass, 10x15 cm in size, compressing the left atrium. a) Parasternal long-axis view, b) Apical four-chamber view.
LA: Left Atrium, LV: Left Ventricle, RA: Right Atrium, RV: Right Ventricle.

DISCUSSION

Desmoid tumours, also known as aggressive fibromatosis, are extremely rare neoplasms first described by MacFarlane in 1831.[1] They arise from musculoaponeurotic tissues and have benign microscopic findings. Their etiology is still unknown. Although these tumours have no metastatic features, they usually have local agressive behavior and surround vital structures such as the aorta, esophagus and trachea, spine and nerves. Radical excision of the tumour is the first choice of treatment and early management is crucial because of potential invasion.[2] Desmoid tumours have intra-

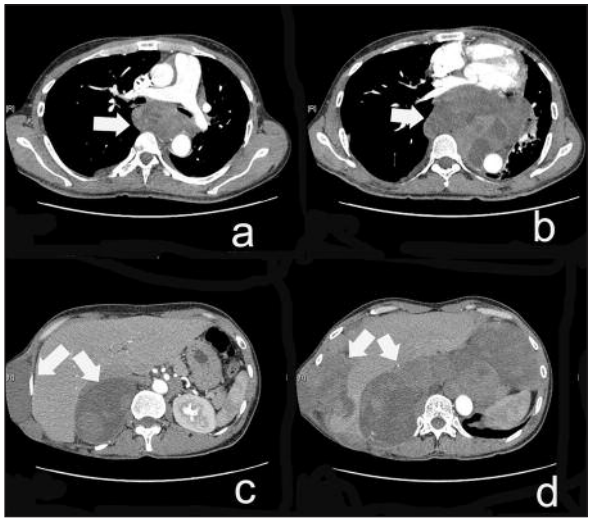

FIGURE 3a-d: Transverse Computed Tomography images displayed desmoid tumour. In upper mediastinum **(a)**; invasion of pulmonary artery, in posterior mediastinum **(b)**; at the back of heart, in retroperitoneum **(c)**; extending to paravertebral column and behind the liver, right lumbocostal region **(d)**. CT also showed invasive appearance of desmoid tumour (arrows).

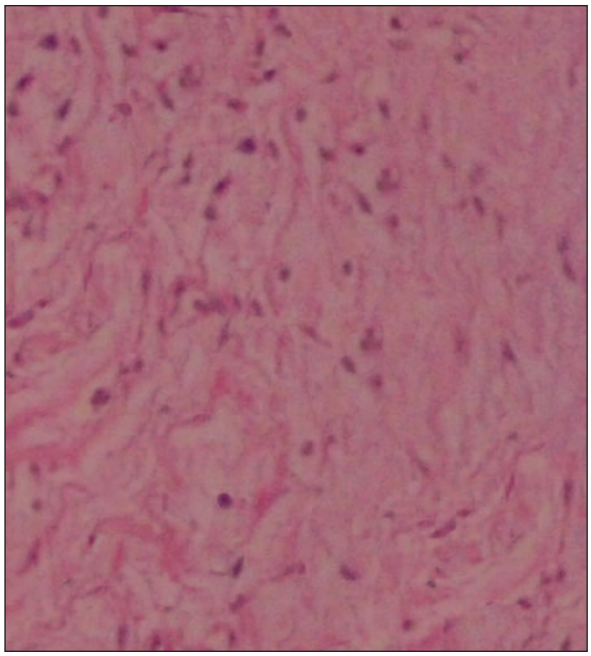

FIGURE 4: Microscopic view of desmoid tumour; Haematoxylin and Eosin (H & E)-stained section revealed typical spindle cells and increased fibroblasts in a collagen matrix (x200).
(See color figure at
http://www.turkiyeklinikleri.com/journal/cardiovascular-sciences/1306-7656/)

abdominal, abdominal and extra-abdominal localizations. The most common placement of desmoid tumour is in the intra-abdominal region.[3] Causative agents are unknown. Limited data is available in the literature about desmoid tumour associated with adenomatous polyposis and Gardner's syndrome. Even when wide margin surgical procedures are performed, very high recurrence rates are seen, such as up to 40%.[4] Though the role of radiotherapy and chemotherapy after surgery remains uncertain, response to radiotherapy seems to be much better than to chemotherapy.[5]

Here, we submit our case of recurrent desmoid tumour with widespread invasion extending from the superior mediastinum to upper retroperitoneum and the right lateral lumbocostal region. As desmoid tumours are locally aggressive, complete surgical resection is the most serious difficulty in treatment. Even though our patient had undergone retroperitoneal surgery, recurrent desmoid tumour advanced covertly over the years and was only noticed by the patient due to a distinct mass in the right lateral lumbocostal skin region in the last six months. As the disease is extremely rare, there is not enough data on its clinical advancement. The surgery records of the patient showed no thoracic involvement at the time of surgery; however, the current involvement extending to the mediastinum and right lumbocostal skin region emphasized that, despite the benign histopathology of the desmoid tumour, it was actually locally aggressive. Transthoracic echocardiogram is a worthwhile inexpensive and noninvasive method used not only in the assessment of cardiac structure and functions, but also in the imaging of the posterior mediastinum. Even though desmoid tumours are very rare occurrences, their high recurrence rates and invasion of vital organs make close follow-up crucial. The failure of the patient to go for regular follow-ups has made it impossible for the desmoid tumour to be diagnosed before invading vital organs and veins. There is no data in the literature on the recurrence of desmoid tumours by advancing from the retroperitoneal region to the superior mediastinum. Also, right lumbocostal involvement in the patient suggested that the cells were planted along the incision line during surgery. The fact that our patient's retroperitoneal desmoid tumour extended covertly

and slowly to the superior mediastinum clearly reveals the value of echocardiographic examination in the follow-up of such cases. Finally, desmoid tumour is an extremely rare disease, but it should be considered among the differential diagnosis of posterior mediastinal and retroperitoneal mass. Though they are mostly localized in the intra-abdominal region, these tumours should be followed-up for involvement of the superior mediastinum by using advanced examinations.

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