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Mesothelial/Monocytic Incidental Cardiac Excrescences (MICE) with Primary Cardiac Leiomyosarcoma: Report of the First Case

Primer Kardiyak Leiomiyosarkom ile Birlikte Olan Mezotelyal/Monositik Rastlantısal Kardiyak Tümör: İlk Vaka Sunumu

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Özet

Nefes darlığı ve bayılma şikayeti ile kardiyoloji polikliniğine başvuran 74 yaşındaki erkek hastanın yapılan transtorasik ekokardiyografisinde sol atriyum içerinde kitle saptandı. Transözefageal ekokardiyografide bir tanesi sol atriyum duvarı ve mitral kapak arka yaprakçığının atriyal yüzüne geniş tabanlı olarak bağlanmış, diğeri ise ilk parçaya bitişik olan 2 parçalı sol atriyal kitle saptandı ve hasta operasyon amaçlı kalp damar cerrahisine devredildi. Cerrahi olarak çıkarılan kitlenin histopatolojik incelemesi mezotelyal/monositik rastlantısal kardiyak tümör (MRKT) ve düşük grade'li leiomiyosarkom olarak rapor edildi. Yapılan literatür incelemesinde, MRKT ve primer kardiyak leiomiyosarkom birlikteliği bildirilmemiştir. Bu yazıda nefes darlığı ve bayılma şikayeti sonrası tanı konulan bu nadir kardiyak kitle ekokardiyografik ve histopatolojik bulguları ile birlikte sunulmuştur.

Anahtar Kelimeler: Kardiyak tümör, kardiyak leiomiyosarkom, sol atriyal kitle, mezotelyal/monositik rastlantısal kardiyak tümör.

Abstract

A 74-year-old man was admitted to outpatient cardiology clinic with the compliant of shortness of breath. After evaluation of the patient, transthorasic echocardiography revealed left atrial mass. Transesophageal echocardiography demonstreated a left atrial mass with two lobule, one attached to left atrial wall and atrial side of posterior mitral leaflet with a wide base, the other one was adjacent to the first one and patient refered to cardiac surgery. Histopathological diagnosis of masses revealed Mesothelial/monocytic incidental cardiac excrescence (MICE) and low grade leiomyosarcoma. According to our search of literature did not yield any report of a case with occurance of primary cardiac leiomyosarcoma MICE together. Thus, we discuss herein this rare cardiac mass presented with shortness of breath and syncope, together with the findings on echocardiography and histopathology.

Keywords: Cardiac tumors, cardiac leiomyosarcoma, left atrial mass, Mesothelial/monocytic incidental cardiac excrescence.

Introduction

Benign and malignant cardiac tumors are rare diseases. Metastatic cardiac malignancies exceed in number primary malignant cardiac tumors by at least a 30 to1 ratio in the majority of autopsy series. Reported incidence of primary cardiac tumors are 0.0017% and 0.03% in consecutive autopsy series (1,2). 25% of all primary tumors of heart is malignant. Angiosarcoma is the most common malignant cardiac tumor. However, the incidence of cardiac leiomyosarcoma is less than 1% (2).

Mesothelial/monocytic incidental cardiac excrescence (MICE) is a rare intracardiac mass which is a mixture of histiocytes, mesothelial cells, fibrin, adipocytes and inflammatory cells and may be a result of inflammation, mechanic irritation or tumor. MICE can be misdiagnosed as cardiac malignencies altough it is considered benign (3).

Present case report represents the togetherness of two rare intracardiac mass; leiomyosarcoma and MICE. To our knowledge this report will be the first case of cooccurrence of leiomyosarcoma and MICE.

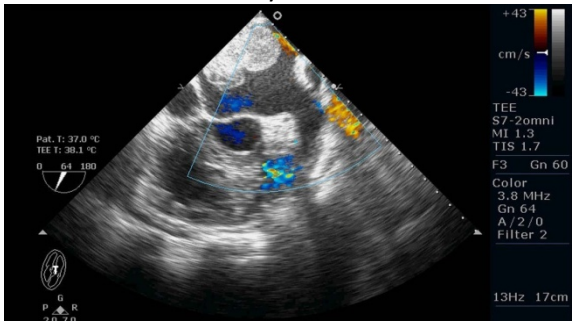

Figure 1. TEE demonstreated a left atrial mass with two lobule.

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Case report

A 74-year-old man was evaluated at outpatient cardiology clinic in another city goverment hospital with the compliant of shortness of breath and syncope. After evaluation, echocardiography revealed left atrial mass and patient referred to our institution for the detection of left atrial mass. Patient admitted to our clinic, on physical examination; blood pressure 100/80 mmHg, pulse 72/min, body temperature 36,7 ºC, respiratory rate 20/min, auscultation revealed 2/6 systolic ejection murmur at apex area of heart. Except elevation of sedimentation rate (72 mm/h) and low hemoglobin (10,2 g/dl) biochemical parameters of blood were normal. Transesophageal echocardiography demonstrated a left atrial mass with two lobule, one attached to left atrial wall and atrial side of posterior mitral leaflet with a wide base, the other one was adjacent to the first one. Mild mitral regurgitation was seen (Figure 1).

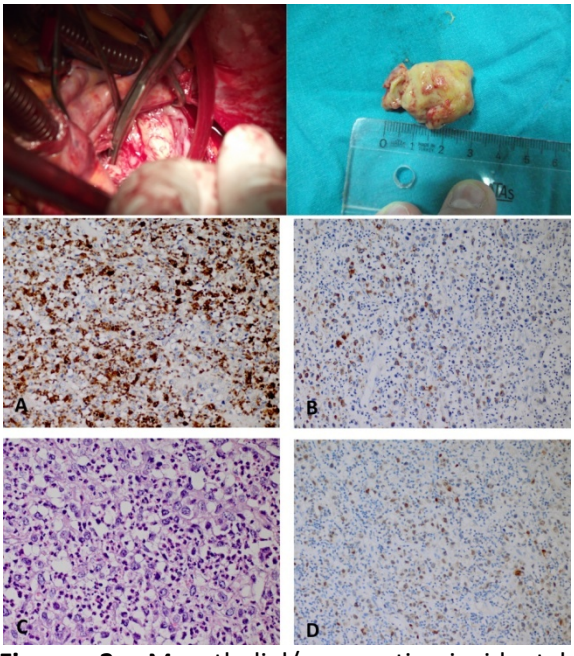

Figure 2. Mesothelial/monocytic incidental cardiac excrescences (MICE); Encapsulated, solid, jellyform appearing as a pale thrombus.(Top left and right) Admixture of mesothelial cells and histiocytes (H&E stainx200)(A), Calretinin-strongly positive tumor cells (x200)(B), Desmin-positive tumor cells (x200)(C) CD 68-positive macrophages (x200)(D)

After coronary angiography, which revealed normal coronary arteries, patient referred to cardiac surgery department and the mass which was attached to left atrial wall and mitral posterior leaflet, resected completely. Histopathological examination of the mass revealed low grade leiomyosarcoma. Light microscopy shows spindle cells with a low rate of mitosis, immunohistochemical staining were negative for CD34, S-100, desmin and positive for actin and focally for myoglobin (Figure 2). Examination of the adjacent mass revealed MICE which was immunohistochemically positive for calretinin and desmin (Figure 3).

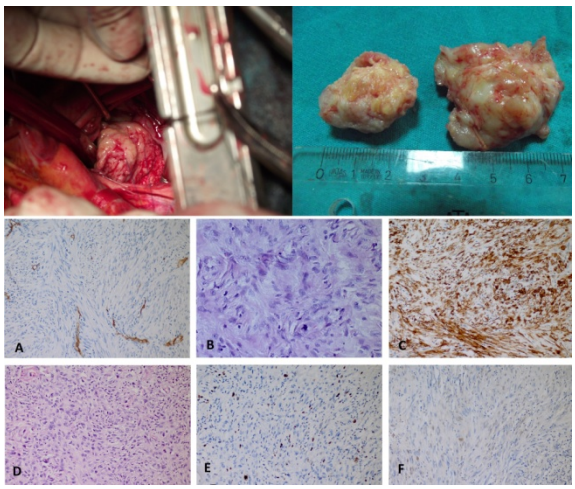

Figure 3. Leiomyosarcoma; seperate, hard, solid, wide based and adjasent to posterior mitral anulus excised by two fragmentated pieces.(Top left and right) İnterlacing fascicles of spindleshaped tumor cells. (H&E stainx200)(A) with markedly atypical nuclei and increased mitotic activity (H&E stainx400)(B), Actin positive tumor cells (x200)(C), Myoglogulin focal positive tumor cells (x200)(D). KI-67 %5-10 nuclear positive tumor cells (x200)(E).CD 34 negative tumor cells (x200)(F).

Discussion

Cardiac tumors may present by dyspnea, pericardial effusions, atrial arrhythmias, congestive heart failure, chest pain and cough. By the advances in diagnostic techniques, it is possible to diagnose the disease when patient is alive. Echocardiography is the most useful tool to detect intracardiac masses. If there is

suspicious, CT and MRI may differentiate tumor and thrombotic material beter (4).

Nearly 25% of primary cardiac tumors are malignant and most of these are sarcomas. These sarcomas include angiosarcomas, leiomyosarcomas, rhabdomyosarcomas, malignant fibrous histiocytomas, undifferentiated sarcomas, fibrosarcomas, and malignant lymphomas. Primary cardiac leiomyosarcoma constitudes a minority of these, less than 1% of all primary cardiac tumors. Approximately 70 to 80 percent of leiomyosarcomas arise from the left atrium and prone to extend into the pulmonary trunk (5-7). Although invasion of mitral leaflet was reported (8), at present case tumor invasion was on left atrial wall, over mitral anulus.

MICE is a benign and rare intracardiac mass, which is a mixture of histiocytes, mesothelial cells, fibrin, adipocytes and inflammatory cells. Exact incidence and etiology of cardiac MICE is unknown, to date fewer than 50 cases have been recorded in the available literature (9). Two theories, 'reactive'' theory and "artifact" theory, have been proposed to explain how the mesothelial cells ''gain'' access into the intravascular compartment. The reactive theory stated that MICE may be a reactive lesion as a result of inflammation, mechanic irritation (cardiac catheterization) or tumor. The artifact theory postulated that MICE are artifactual, formed by the manipulation of the cardiac surgeon, compacted by suction vacuum at the time of the procedure (3,10). In our case, no prior cardiac catheterization or cardiac surgery was performed. To the best of our knowledge, there has been no report of a case with occurrance of primary cardiac leiomyosarcoma and MICE together. Furthermore present case may support the reactive hypothesis about the etiology of MICE.

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