INTRODUCTION

Vascular malformations (VMs) of the heart are extremely rare with only a few cases of the arteriovenous type of vascular malformation (AVM) reported (1,2,3,4,7,9). The frequency of primary cardiac tumors seen at autopsy is ≈0.02%. Although cardiac lymphangiomas are often asymptomatic, the main symptoms include dyspnea, palpitation, atypical chest pain, and arrhythmia. Echocardiography represents the diagnostic imaging modality of choice to appropriately screen for cardiac tumors. Computed tomography and magnetic resonance imaging are complementary. Surgical resection is the treatment of choice for symptomatic cardiac lymphangioma and is considered to be curative in most cases. (8,10,11)

CASE REPORT

A 28-year old woman was admitted to the hospital with symptoms of exsional dyspnea. Physical examination was normal except a 2/6 systolic murmur. Chest x ray and ECG was normal.

Trans thoracic and transesophageal echocardiography showed a large mass (4.2 X 3.5cm) in the lateral side of right atrium (RV) and atrioventricular (AV) groove with extension to RV outflow tract (RVOT) without obstruction and mild PI (figure 1). Other valves and chambers of heart were normal.

Abstract: Cardiac lymphangioma are rarely seen in young adult with atypical presentation. Here in we presented a 28-year-old woman with atypical presentation of large cardiac lymphangioma encased RCA and Pulmonary artery and was inoperable.

Key word: Lymphangioma; Cardiac Tumor; Atypical Presentation

Figure 1: Echocardioghapic pictures of right ventricle lymphahgioma.
Cardiac MRI showed a large intra-cavitary RV mass (85 X 43 mm) which encased the (Right coronary artery) RCA with involvement of RV anterior wall, right AV inflow, encircling RVOT with extension to pulmonic annulus and aortic root (figure 2).

Pathologic examination revealed proliferation of dilated vascular channels without any content with thin wall lined by flat endothelial cells in background of edematous fibrous stroma and focal mature lymphocytes aggregation suggestive of lymphangima (figure 4).

Figure 2: Cardiac MRI (T2) Of Right Ventricle

Coronary angiography showed a normal coronary artery without vascular origin. Based on the suspicion of malignancy (angiosarcoma), cardiac biopsy was planned. After median sternotomy my finding a cluster of small vessels on the surface of RV and pulmonary valve (figure 3). Two big specimens (2*2cm) were taken and sent for frozen section and pathologic examination. Frozen section showed vascular malformation without any finding of malignancy. The patient was discharged six days post operation.

Figure 3: Patghologic view of of right ventricle lymphanghioma.

At two year follow up the patient with echocardiography was alive and with mild exsional dyspnea.

DISCUSSION
Cardiac lymphangioma is a very rare tumor of the heart, which first reported in 1911 by Armstrong and Monck berg (12). Only nine cases of cardiac lymphangioma have been reported in the medical literature (4,6,7,8). More than half of the reported cases have occurred in patients under 10 years of age (8). Mediastinal lymphangiomas are most often asymptomatic masses incidentally discovered on chest X-ray (9). The tumors most commonly occur in the pericardial space, but other unusual primary sites include the myocardium, the posterior wall of the left atrium, and the AV node regions (10). The primary site of the cardiac lymphangioma in the presented case (lower part of the interatrial septum) differs from all prior reports of this tumor (11). Cardiac lymphangioma can be successfully excised, and surgical resection is the treatment of choice for symptomatic lesions or when diagnosis is in question. The long-term outcome of patients with surgically treated symptomatic lesions is excellent. Therefore, surgical excision may be unnecessary, particularly for extensive type or asymptomatic lymphangioma.

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