sons, it is unclear that how interpret APA levels in APS. Also Rodriguez-Garcia et al. (5) showed that there is no differences between seronegative and seropositive APS in terms of thrombotic events. A case report which was published by Middlebrooks et al. (6) there is an association between temporary APA positivity and recurrent stent thrombosis. This case was explained by seronegative APS or long-term antithrombotic therapy by the authours. Additionally it was shown that especially aspirin can reduce APA levels (7). In our patient, there is no other possible risk factors responsible for shunt thrombosis except positive APA levels. Based on publications mentioned above we affirm that mBT shunt thrombosis might be related to positive APA levels in this case.

Certain thromboembolic factors such as protein C deficiency, factor V Leiden or protrombin 20210 mutation have been reported with cardiac thrombosis in children (8, 9). Deally et al. (10) published a newborn with mBT shunt thrombosis caused by APS in 1999. This case has antithrombin III deficiency, also. However, there was only one trombophilic factor in our patient. He is the first case in the literature in this regard.

Conclusion

Especially in patients with recurrent shunt thrombosis, hereditary thrombophilia should be investigated and APA levels should be measured in these patients.

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A very rarely seen cardiac mass (Rosai-Dorfman disease)

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Introduction

Sinus histiocytosis (Rosai-Dorfman Disease) is a rare disease, which is characterized by massive lympadenopathies with unknown etiology. It was first defined in 1969 by Rosai and Dorfman (1). Although it is seen most frequently in the first two decades of life, it can be observed at any age. The frequency of cardiac involvement is less than 1% in Rosai-Dorfman disease (RDD) (2). Here, we report a case with extranodal RDD in which cardiac involvement was detected.

Case Report

A 62-years-old male patient was referred to cardiology clinic with the complaints of atypical chest pain and dyspnea. His physical examination was unremarkable. A cardiac mass with 2x1.8 cm dimentions attached to wall of the right atrium was observed in transthoracic echocardiography (Fig. 1). Thoracic computerized tomography (CT) (Fig. 2) and Cardiac Magnetic Rezonans (MRI) showed a mass with 37x29 mm dimensions originating from the wall of the superior vena cava and extending to the interatrial septum, and along the lateral right atrial wall to the atrioventricular groove (Fig. 3A, B).

Surgical technique

Under general anesthesia, median sternotomy, standart aortic cannulation and selective bicaval cannulation were performed. A gray-

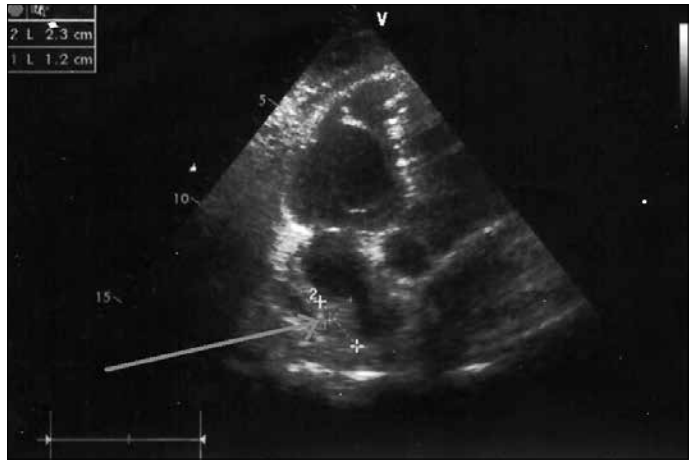

Figure 1. Showing a right atrial mass in Echocardiography

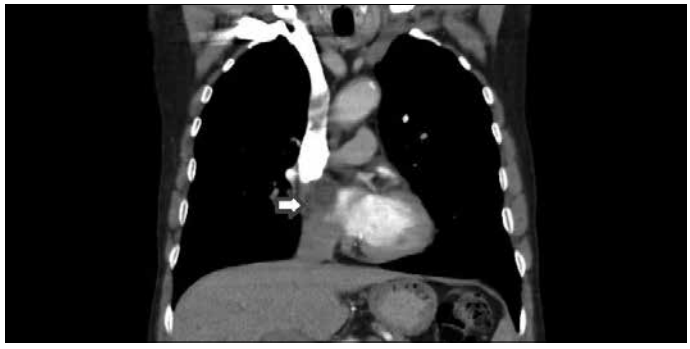

Figure 2. Showing a right atrial mass in thoracic computed tomography

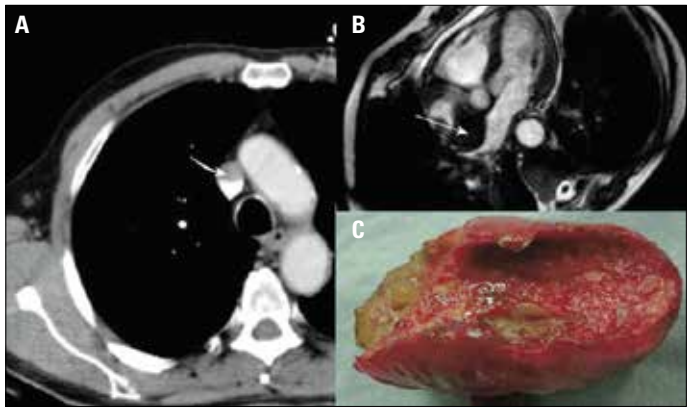

Figure 3. (A) The computed tomography image of the mass narrowing the ostium of superior vena cava. (B) Cardiac MR showing a right atrial mass. (C) Macroscopic view showing a cardiac mass

yellow coloured solid mass, located on the cavo-atrial junction was seen during surgical exploration. (Fig. 3C). The mass was excised subtotally because the invasion to the interatrial septum and the wall the of right ventricule made a total excision irreparable. We aimed to remove the hemodynamically significant part of the mass. Postoperative follow up was uneventful and he was discharged from hospital at postoperative 7th day.

Postoperative immunohistochemical studies showed diffuse infiltrations of hystiocytic cells between cardiac muscle cells with lymphocytes, fibroblast*s* and perivascular myxoid degeneration (Fig. 4). In the histopathologic examination phagocytosis of lymphocytes and plasma cells by some hystiocytes (emperipolesis) is seen (This hystiocytic cells were positive for CD68, S100 and CD 163 proteins and they were negative for langerin and CD1a) (Fig. 5).

Discussion

RDD is a rare multisystem disorder. The common symptoms include painless, bilateral cervical lymphadenopathy with fever**.** Diagnosis of RDD is only made by histopathologic examination. The most important histopathologic feature of this disease is emperipolesis (lymphophagocytosis). Emperipolesis is the detection of lymphocytes and sometimes plasma cells and erytrocytes in histiocytes (3).

In our case, depending upon the morphologic and immuno-histochemical findings, RDD was diagnosed. That may be the proof of extranodal RDD disease. Cardiac involvement can be seen in all chambers of heart, valves, pericardium, aorta and pulmonary arteries (4). Ajise et al. (5) reported that chest pain, dyspnea, atrial fibrillation and hypotension were seen in the 61-year-old patient who had RDD in right

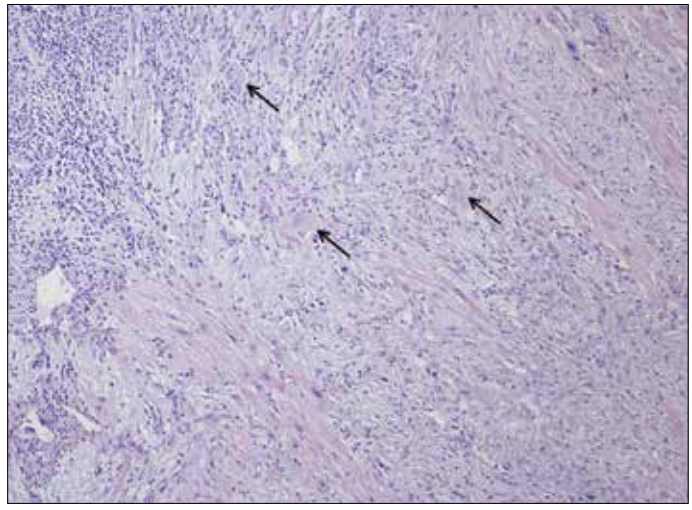

Figure 4. Large histiocytes with eosinophilic cytoplasm and some multinucleated giant cells originating from them (arrow) are seen between muscle cells on an inflammatory ground rich of lymphocytes. (H/E - original magnification: x10)

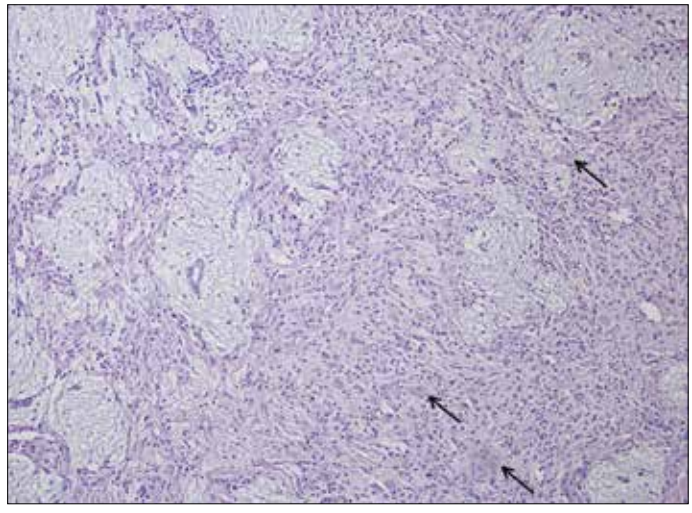

Figure 5. Lymphocytes phagocytized by histiocytes (emperipolesis). (H/E - original magnification: x10)

atrium. Richter et al. (6), reported that they operated a patient with RDD in left atrium and ventricle who suffered from atypical chest pain.

Also, Maleszewski et al. (7) , reported two patients with RDD; one of them (40-year-old male) who had right atrium involvement was diagnosed accidentally and the other was a 57 years old female with the complaint of chest pain. Recently Sarraj et al. (8), reported a case with RDD in left ventricle anterolateral region (29-year-old male) with complaints of palpitation. They did not detect any skin lesions or cervical lymphadenopathies. In our case RDD was located in right atrium and narrowed the opening of SVC.

Indications of surgical treatment are compression to the vital organs, cosmetic deformity, situations that lead functional disorders and life threatening conditions (9).

Cases with RDD without extranodal involvement have a benign clinical progress usually and clinic follow up is enough in those cases. Aggressive behavior and mortality is rarely seen. RDS with extranodal involvement has poor prognosis, especially when liver, kidney and respiratory tract are involved (10). Our case was different than other cases because patient's mass located in the right atrium caused

significant narrowing at the entrance of superior vena cava. Treatment options may differ from case to case depending upon the location of extranodal RDD. Our case needed cardiac surgery because of the narrowing of SVC opening and coronary artery disease.

Conclusion

As a result, we think that RDD must be kept in mind in the differential diagnosis of cardiac masses. There may be some difficulties in diagnosis without getting tissue sample in RDD. Cardiac involvement needs more aggressive treatment because it may cause some life-threatening conditions.

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