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Cardiology

A Rare Case of Left Atrial Myxoma Associated with Behcet's Disease

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Abstract **Case Report**

Behcet's disease is a rare form of auto immune multisystemic disease also called as auto immune vasculitis. The three cardinal features of Behcet's disease are oral ulcers, genital ulcers and uveitis. Diagnosis is based on clinical, radiological and laboratory investigations. In the absence of constitutional symptoms, diagnosis of Behcet's disease could be challenging. Cardiac involvement of Behcet's disease is extremely rare and can present as intracardiac mass or thrombus with greater predisposition towards right side chambers. Sometimes, the cardiac manifestations could be the presenting symptoms even before the symptoms of Behcet's disease start to appear.

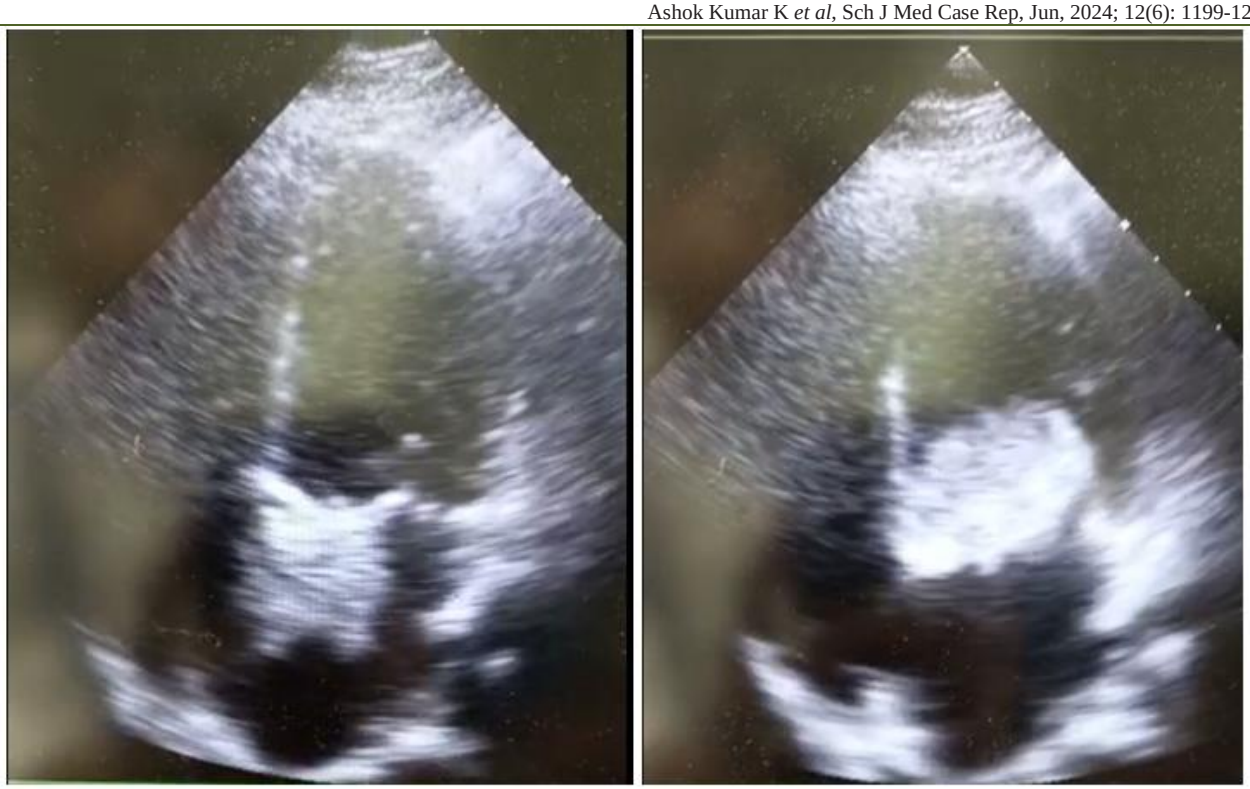
Keywords: Behcet's disease, Myxoma, vasculitis.

INTRODUCTION

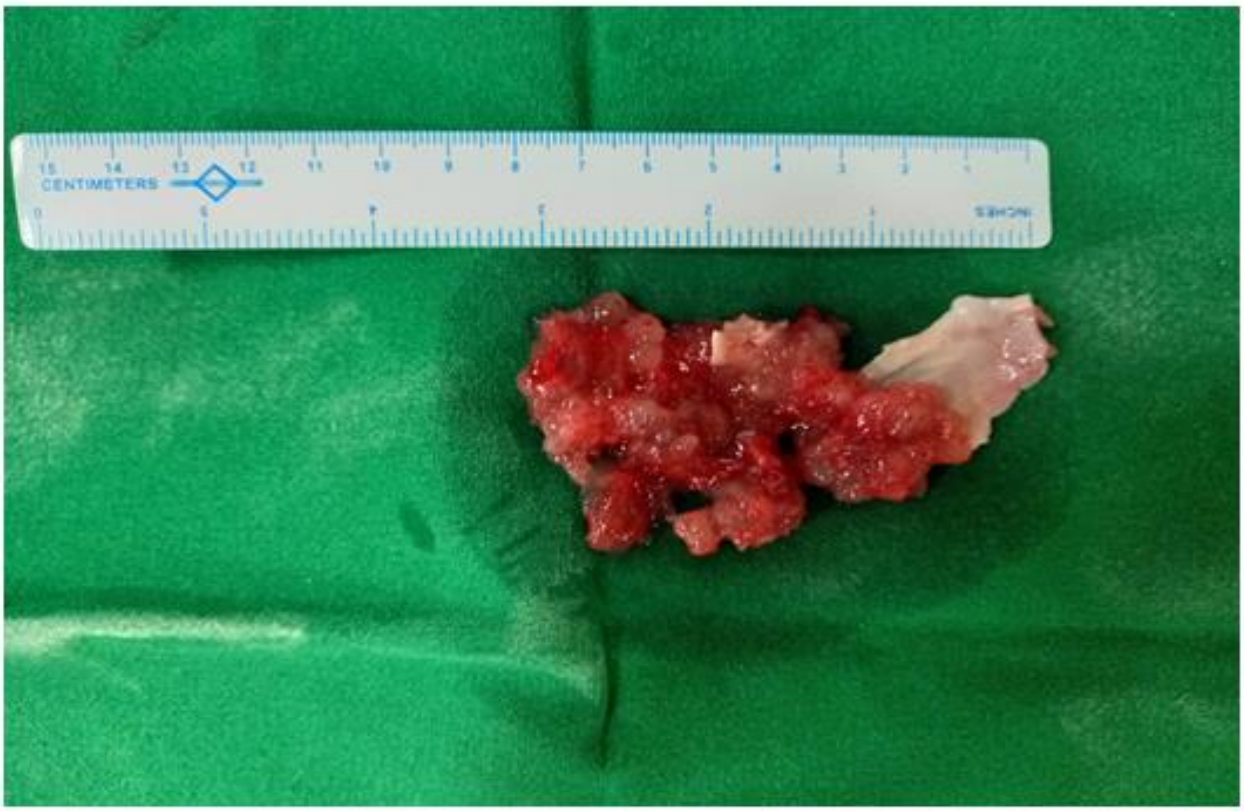
Behcet's disease affects predominantly female population between 20 to 40 years age group and mostly seen in the mediterranean region. The commonest cardiac manifestation of Behcet's disease is an intra cardiac mass or thrombus, more commonly in the right ventricle followed by the right atrium. Left sided cardiac chamber involvement in Behcet's disease is extremely rare. In this case report, we present a rare combination of Behcet's disease with left atrial myxoma. In the absence of constitutional symptoms, diagnosis of Behcet's disease could be challenging.

CASE REPORT

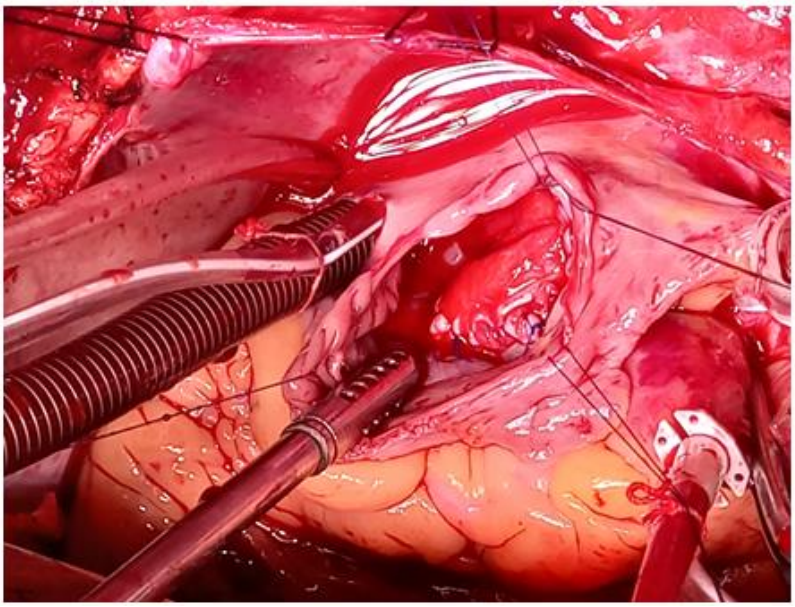
A 40 year old female patient came with complaints of dyspnoea and occasional palpitations since 3 months. She had an episode of cerebrovascular accident with left hemiparesis a year before and was treated for the same. During this period, she noticed painful skin lesions in her feet and hands. After thorough evaluation for various autoimmune/immunological disorders, it was deduced to be Behcet's disease and she was started on a course of steroids. Her symptoms improved gradually. However, during evaluation, 2D echocardiography revealed a large left atrial mass measuring 4.7cm x 3 cm with its stalk attached to the lower part of the inter atrial septum, protruding through the mitral valve into the left ventricle. Mild mitral regurgitation was also seen. It was decided to excise the mass. Under cardiopulmonary bypass, right atrium was opened to approach the myxoma. It was a large, friable, left atrial myxoma attached to the inter atrial septum at the lowermost aspect abutting the wall of the coronary sinus. Through inter atrial septum, total excision of the myxoma was done ensuring that the entire mass along with the stalk and the adjacent septum was excised. Mitral valve leaflets were tested with saline insufflation and were found to be competent. The inter atrial septum was closed using autologous pericardial patch. Right atrium was closed with 5-0 polypropylene sutures in two layers. Coming off bypass was uneventful. Serial decannulation was done. After complete haemostasis, chest was closed and patient was shifted to recovery with stable haemodynamics. Post operative period was uneventful. The specimen was sent for histo-pathological examination. Post-operative 2D echocardiography showed no residual myxoma, intact septum and no mitral regurgitation. Patient was discharged on 4th post op day with stable vitals and prescribed a course of steroids. Histopathological features were consistent with left atrial myxoma.



2D Echocardiography showing a large left atrial mass arising from inter-atrial septum



Excised mass in toto



Closure of the inter-atrial septum with autologous pericardial patch

DISCUSSION

Behcet's disease is a rare form of multi-systemic vasculitis that affects both arteries and veins and it can have frequent relapses. Though the three cardinal features of Behcet's disease are uveitis, oral ulcers and genital ulcers, it can manifest in other systems too including central nervous system, cardiovascular and gastrointestinal system. There is no one set of investigations specific for Behcet's disease though there may be increased inflammatory markers. The commonest cardiac manifestation of Behcet's disease is an intracardiac thrombus with more predisposition towards the right sided chambers. It is unclear why the thrombi in Behcet's disease have increased propensity for right sided chambers [1]. Diagnosis of intra cardiac mass is extremely challenging; being mandatory in distinguishing between thrombus, neoplasm and vegetation [2]. Transthoracic echocardiography is gold standard in identifying intracardiac mass in Behcet's disease [3]. The exact cause of thrombus formation in Behcet's disease is not well understood. The probable cause might be chronic inflammation of the endocardium leading to endomyocardial fibrosis similar to that which is seen in vasculitis.

Cardiac manifestation in Behcet's disease may be in the form of intracardiac thrombus, rupture of sinus of valsalva, pericarditis, pseudoaneurysm, cardiomyopathy, myocarditis, endocarditis, valvular dysfunction or rupture of the sinus of Valsalva [2,3]. In an analysis of 25 patients by Mogulkoc N *et al*., more than half were found to have thrombus in right ventricle (52%). Right atrium was second most commonly involved cavity (24%). Left atrium and left ventricle were least affected chambers (4% each) and multi chamber thrombi were found in 16% of the patients [4].

CONCLUSION

Left atrial myxoma could be a rare association of Behcet's disease. In the presence of intra cardiac mass with constitutional symptoms like fever, weight loss or breathlessness, Behcet's disease could be one of the possible diagnosis, especially if the patient is a young female from mediterranean region or middle east Asia. Behcet's disease associated with left atrial myxoma has not been reported till date. Our closest reference was a left atrial thrombus mimicking myxoma which was documented by WY Madanat *et al*., We are herewith presenting this case report as it would be of interest to both physicians and surgeons.

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