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MYXOMAS OF THE LEFT ATRIUM AND LEFT VENTRICLE WITH COEXISTENT RHEUMATIC DISEASE OF THE MITRAL VALVE.

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ABSTRACT

We report a case of 20year young female with shortness of breath, hemoptysis, left sided chest pain and generalized weakness. Transthoracic echocardiography showed multiple soft homogenous growth in left atrium and ventricle involving the interventrilcular septum, posterior and lateral walls, and left ventricular apex extending to the anterior mitral leaflet with moderate to severe mitral regurgitation. The per-operative findings were a lobulated mass mainly in the left ventricular cavity involving both leaflets of mitral valve and the area adjacent to the medial commissure of the mitral valve in the left atrium. Excision biopsy of the mass and excised mitral valve showed myxoma of left atrium and ventricle with rheumatic disease of the mitral valve. This is a rare case of cardiac tumor presenting as a left ventricular mass along with co-existent rheumatic valve disease.

Key Words: Cardiac tumor, myxoma of left ventricle.

INTRODUCTION

Cardiac myxomas are rare tumors which may present in many different ways. Seventy five percent of myxomas are found in the LA, but 10–20% arise in the right atrium, and 5% in either ventricle. Left ventricular myxoma is a rare benign cardiac tumor. Surgical excision is the treatment of choice and completeness of removal is mandatory to avoid late recurrence[1,2]. Patients with myxomas who become symptomatic usually present with congestive heart failure (67%) because of LV inflow obstruction, signs of tumor embolization (29%), or constitutional symptoms (41%).

This report therefore concerns about the clinical and the diagnostic approach of an atypical case of cardiac myxoma. In addition, a short review of literature is included.

CASE REPORT

A 20-year-young female presented in the outpatients with shortness of breath, hemoptysis, left sided chest pain and generalized weakness for the past one year. She could not walk and was wheelchair dependant when she arrived for consultation. On auscultation there was a pansystolic murmur at apex radiating to the axilla. Chest x-ray was normal.

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Transthoracic echocardiography demonstrated multiple soft homogenous growths along the basal-inferior interventriclular septum, extending to the anterior mitral leaflet causing both left ventricular inflow and outflow obstruction (fig-1).

The anterior mitral leaflet was found to be prolapsing. There was moderate to severe mitral regurgitation with enlargement of the left atrium (LA 58mm). The aortic valve was normal as was left ventricular function, while the right ventricle was enlarged and hypokinetic (fig-2). The tricuspid valve was normal in size and function. Preoperative transesophageal echocardiography (TEE) demonstrated a 2 x 2 cm LV mass with multiple masses on the septum and along left ventricular inflow and outflow tracts involving the posterior as well as lateral walls and the left ventricular apex. A CT Scan confirmed the diagnosis corroborating the Echocardiographic findings (fig-3).

There was a history of acute throat infection in childhood with knee joint pain occurring thereafter. This case was presented in the weekly hospital cardiology/cardiac surgical meeting and as these masses were widespread, an extensive resection of the septum and inflow and outflow tracts was thought to be pointless. As surgery for excision of the tumor has been discounted, a mitral valve

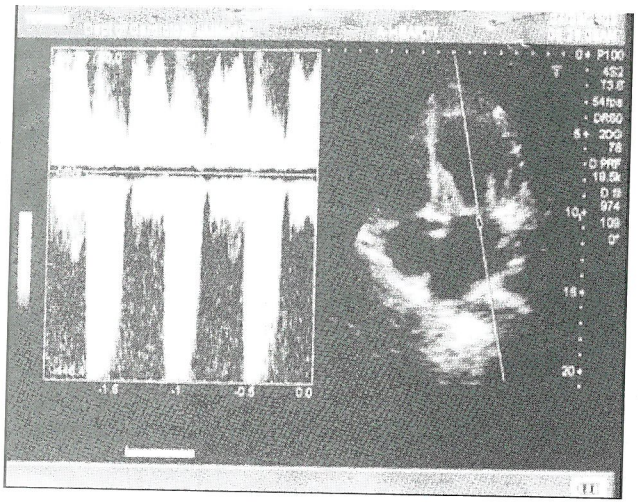

Fig-1:
Pre-operative Mitral Stenosis and Regurgitation

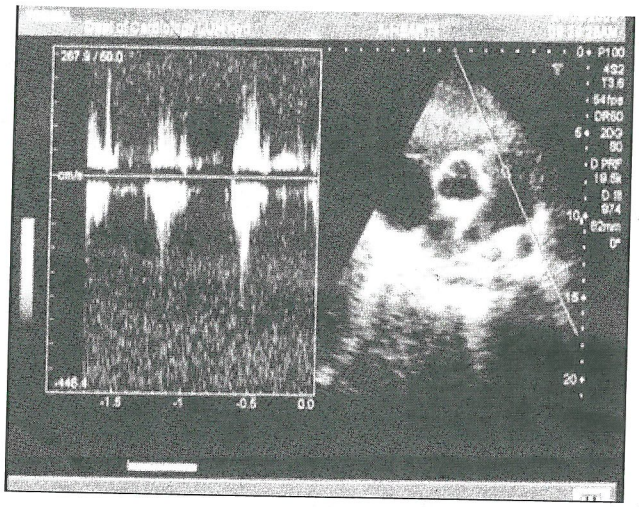

Fig-2:
Pressure gradients across the RV.

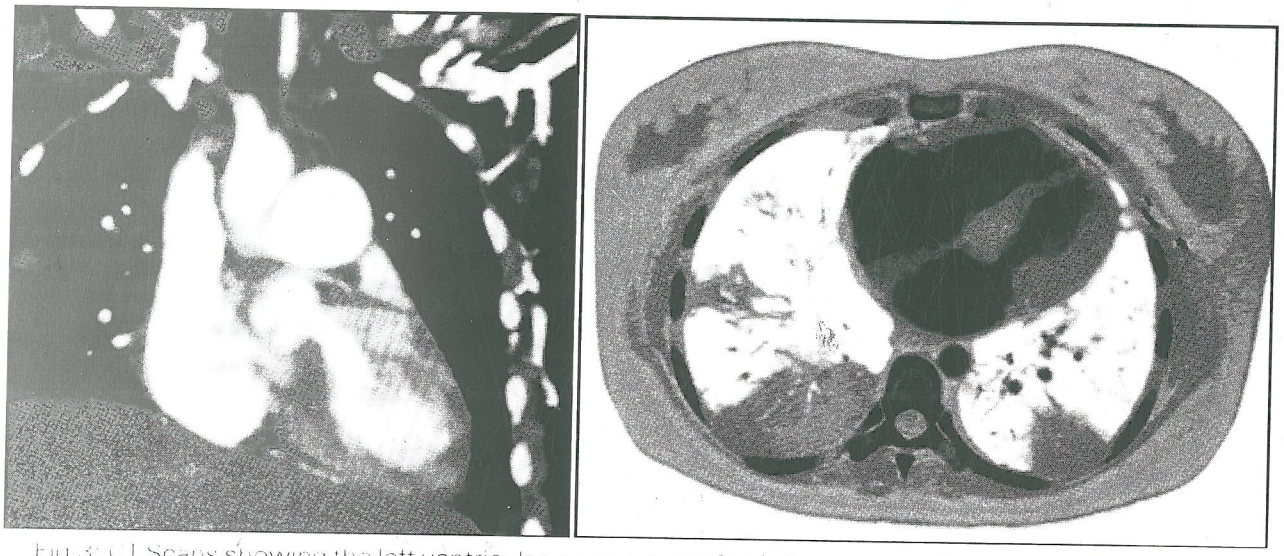

Fig 3: CT Scans showing the left ventricular myxoma causing inflow obstruction and thickening of the ventricular septum and basal walls.

replacement was planned in view of the moderate MR with the object of relieving her symptoms to some extent and obtaining tissue diagnosis at the same time. Standard midline sternotomy incision was fashioned. The pericardium was opened using an inverted T-shaped incision. Heparin (50 U/kg) was given to achieve anticoagulation. The activated clotting time (ACT) was maintained at more than 480 seconds. With ascending aortic return, bicaval atrial cannulation was performed to achieve cardiopulmonary bypass. The aorta was cross clamped and myocardial protection was achieved with intermittent antegrade cold blood cardioplegia (4:1 blood to crystalloid ratio) and moderate systemic hypothermia (28C°). A left atriotomy was performed to gain access to the mitral valve. A small mass was found adjacent to the medial commissure and on retraction of the anterior mitral leaflet, further masses were found along the septum and lateral wall. These masses were peeled and teased off the ventricular walls with blunt and sharp dissection, and removed in toto. A thorough inspection was carried out for presence of other tumors. Following this, the area was extensively washed and suctioned. Mitral Valve Replacement was carried out with a size 29 Carbomedics bileaflet prosthesis. The left atriotomy was then closed, with left atrial venting, as the heart slowly assumed sinus rhythm. Inotropic support was started before separation from bypass. Intra-operative TEE confirmed complete excision of the mass.

Histopathologic examination confirmed the mass to be a myxoma. Post-operative recov-

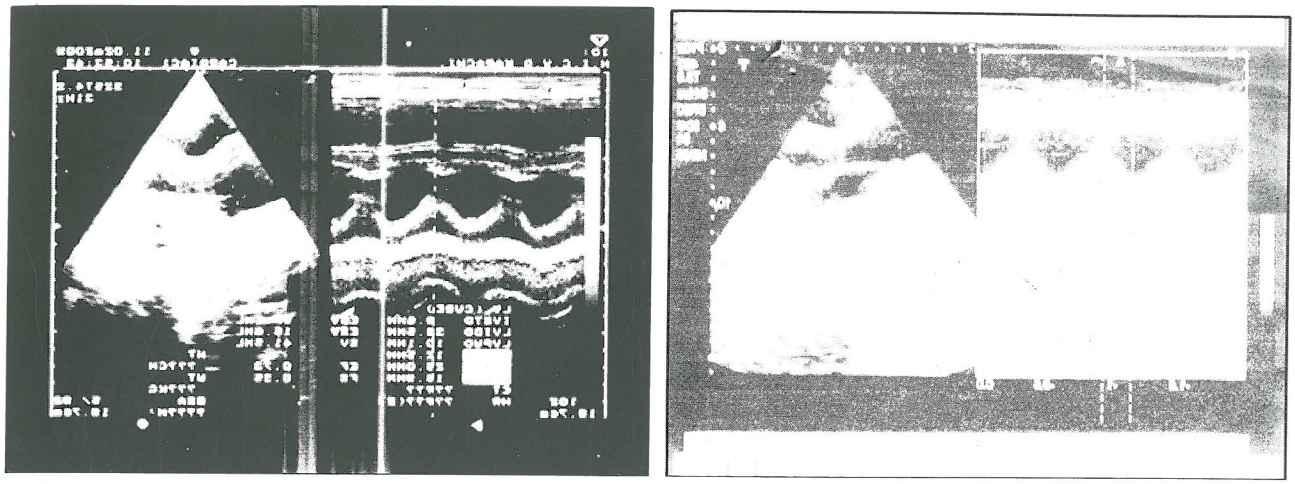

Fig-4: Preoperative and postoperative left ventricular dimensions showing complete excision of tumour.

ery was uneventful and the patient was discharged home on the 10th postoperative day after satisfactory anti-coagulation was confirmed. Monthly follow-up achieved with thorough clinical examination and serial echocardiograms (fig-4). The patient was discharged from out-patients after her third follow-up visit and will be reviewed after one year.

DISCUSSION

Neoplasms of the heart can be primary cardiac tumors arising in the heart and secondary cardiac tumors that have metastasized to the heart. Primary tumors of the heart are rare[1,2] .Primary cardiac tumors can be benign and malignant tumors.[7] Approximately 50% of the benign tumors are myxomas.

Myxomas occur in any chamber of the heart but most commonly from left atrium (75%) and the right atrium (10%–20%). The remaining 6% to 8% are equally distributed between left and right ventricles.[1,2,7,11] Majority of myxomas occur sporadically, more commonly in women than men. The peak incidence is between the third and sixth decades of life (middle-aged) and 94% of tumors are solitary. About 5% of myxoma patients show a familial tendency based on autosomal dominance inheritance, commonly in younger patients with equal incidence in male and female, and more often (22%) have multi-centric disease arising from the atrium or ventricle, the cardiac valves, pulmonary artery and vein, and vena cava.[7,13] Familial myxoma has a higher recurrence rate after surgical resection (21%–67%).

Ventricular myxomas occur more commonly in women and children and may be multi-centric. Right ventricular tumors arise from the free wall while left ventricular tumors tend to occur in the proximity of the posterior papillary muscle[7].

Most myxoma tumors grow rapidly, but growth rates vary and occasionally tumor growth arrests spontaneously.

The classical triad of myxoma presentation is intra-cardiac obstruction with congestive heart failure (67%), signs of embolization (29%), systemic or constitutional symptoms[3,4,5] of fever (19%), weight loss or fatigue (17%), lethargy and immunologic manifestations of myalgia, weakness, and arthralgia (5%). Cardiac rhythm disturbances and infection are rare. These symptoms often suggest an inflammatory autoimmune disease and are unrelated to the location and size of the tumor. There may be leukocytosis, elevated erythrocyte levels and sedimentation rate, hemolytic anemia, thrombocytopenia, and elevated C-reactive protein. Immuno-electrophoresis may reveal abnormal immunoglobulin levels with increased circulating IgG. Interleukin-6 level is also elevated in patients with myxoma suggesting a link to a variety of conditions including lymphadenopathy, tumor metastasis, ventricular hypertrophy, and development of constitutional symptoms.[8,9,15]

Raynaud's phenomenon, arthralgias, myalgias, erythematous rash, and clubbing of the digits are other less frequent complaints. Possible etiologies of such complaints and symptoms include tumor embolization with secondary myalgias and arthralgias and elevated immunoglobulin response. Circulating

antibody tumor antigen complexes with complement activation may also play a role in the constitutional symptom. Such symptoms resolve following surgical resection of tumor.

Obstruction of blood flow in the heart is the most common cause of acute presenting symptoms. The nature of these symptoms is determined by which of the chambers is involved and the size of the tumor. Large ventricular myxomas may present as ventricular outflow obstruction and may produce subaortic or aortic valvular stenosis.

Systemic embolization is the second most common mode of presentation for patients with myxoma (30% to 40%). Because the majority of myxomas are left-sided, approximately 50% of embolic episodes affect the central nervous system owing to both intracranial and extracranial vascular obstruction. The neurologic deficits following embolization range from transient to permanent. Specific central nervous system consequences include intracranial aneurysms, seizures, hemiparesis, and brain necrosis. Retinal artery embolization with visual loss can also occur. Infection arising in a myxoma is a rare complication and produces a clinical picture of infectious endocarditis.

Clinical examination findings at the time of clinical assessment of a patient with cardiac myxoma vary according to the size, location, and mobility of the tumor. The murmur of cardiac myxoma may depend on its position. Echocardiography (trans-esophageal) is the investigation required for diagnosis of myxoma. Intraoperative TEE is used for identifying tumor and to confirm the complete surgical excision of a myxoma.[6] Three-dimensional TEE may be helpful to delineate the extent of the myxoma's attachment, so more accurate surgical planning can achieved.

Both CT and MRI detect tumors as small as 0.5 to 1.0 cm but both are used for malignant tumors of the heart. Computerised tomography (CT) or MRI should be reserved for the situation in which the diagnosis or characterization of the tumor is unclear after complete echocardiographic evaluation.

Surgical excision is the treatment of choice and should not be delayed because death from obstruction to flow within the heart or embolization may occur and completeness of removal is mandatory to avoid late recurrence.[1,2,14,16]

Standard cardiopulmonary bypass is established with ascending aorta and bicaval cannulation. Manipulation of the heart before initiation of cardiopulmonary bypass should be minimal because of the friability and embolic tendency of myxomas. Because myxomas rarely extend deep in the endocardium, it is not necessary to resect deeply around the conduction tissue. Regardless of the surgical approach, the resection should include the tumor and a portion of the cardiac wall to which it is attached. Ventricular myxomas usually are approached through the AV valve or by detaching the anterior portion of the AV valve for exposure and resection and reattachment after resection.[7]

It is not necessary to remove the full thickness of the ventricular wall since no recurrences have been reported with partial thickness excisions. As with right atrial myxoma, the presence of ventricular myxoma prompts inspection for other tumors because of the high incidence of multiple tumors. Try to remove the tumor without fragmentation. Following tumor removal from the field, the area should be liberally irrigated, suctioned, and inspected for loose fragments.[7]

Minimally invasive approaches can be used for cardiac tumors excision. Approaches have included right parasternal or partial sternotomy exposure with standard cardioplegic techniques, right submammary incision with femoral-femoral bypass and non-clamped ventricular fibrillation,and right submammary port-access method with antegrade cardioplegia and ascending aortic balloon occlusion. Results are good but more experience and longer follow-up are needed before this can be recommended as a standard approach.

About two thirds of myxomas are round or oval with a smooth or slightly lobulated surface, polypoid, relatively compact, pedunculated, mobile, and not likely to fragment spontaneously. Mobility depends on the length of the stalk, the extent of attachment to the heart, and the amount of collagen in the tumor. Most tumors are pedunculated with a short broad base. Villous or papillary myxomas may be gelatinous and fragile and prone to fragmentation and embolization. Myxomas are white,

yellowish or brownish color, frequently covered with thrombus. Focal areas of hemorrhage, cyst formation, or necrosis may be seen in cut section. The average size is about 5 cm in diameter but sometimes they can present as a larger mass. Myxomas arise from the endocardium. Histologically, they are composed of polygonal shaped cells and capillary channels within an acid mucopolysaccharide matrix. The base of the tumor contains a large artery and veins that connect with the subendocardium but do not extend deeply beyond the subendocardium in most cases.

A study of last 15 years with 85 myxomas shows no operative or hospital mortality. Recurrence of non-familial sporadic myxoma is approximately 1% to 4%. It probably is even lower in patients with a normal DNA genotype. Twenty percent of patients with sporadic myxoma and abnormal DNA have a recurrence rate between 12% and 40%. The recurrence rate is highest in patients with familial complex myxomas, all of whom exhibit DNA mutation, and this is estimated to be about 22%. Overall, recurrences are more common in younger patients. The disease-free interval averages about 4 years and can be as brief as 6 months. Most recurrent myxomas occur within the heart, in the same or different cardiac chambers, and may be multiple. Relationship of local recurrence to the adequacy of the original resection remains unsettled because sporadic tumors rarely recur even if full-thickness excision of the base is not done and because recurrent tumors often do not recur at the site of the original tumor. Extracardiac recurrence after resection of tumor, presumably from embolization and subsequent tumor growth and local invasion, has been observed. The biology of the tumor, dictated by gene expression rather than histology, may be the only reliable factor predicting recurrence. DNA testing of all patients with cardiac myxoma may prove to be the best predictor of the likelihood of recurrence.[7,16,18]

Patients should be closely followed who are treated initially for multi-centric tumors, those whose tumors are removed from unusual locations in the heart, incompletely resected tumor, and tumors found to have an abnormal DNA genotype and patients undergoing resection of tumors thought to be myxoma but with malignant characteristics at pathologic examination should have long-term, careful follow-up.

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