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LEFT ATRIAL MYXOMA CAUSING FUNCTIONAL MITRAL STENOSIS, MASKED MITRAL REGURGITATION AND DILATATION OF MITRAL VALVE ANNULUS –CASE REPORT

Cardiology

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ABSTRACT

Left atrial myxomas are common benign tumor of heart, it produces various intra-cardiac pathologies which can be missed on 2D Echocardiography and cardiac MRI. we present a case of left atrial myxoma causing functional mitral stenosis, masked mitral regurgitation and dilatation of mitral valve annulus.

KEYWORDS

Left Atrial Myxoma, Mitral Stenosis, Mitral Regurgitation

INTRODUCTION

Myxomas are primary benign tumors of heart having incidence rate 0.5%(1,2), commonest is left atrial(LA) myxoma originating near fossa ovalis of inter atrial septum(1). Myxomas are usually pedunculated and polypoidal(1,2), rarerly due to mass effect they can obstruct blood flow across mitral valve. Vital role in mitral valve leaflets coaptation is played by mitral valve annulas(3). Mitral annular dysfunction can be functional, or due to other disorders like ischemic mitral regurgitation, mitral valve prolapse, atrial fibrillation. Significant dilation of mitral annulas due to large atrial myxoma is rare(4). Here we present case of LA myxoma with functional mitral stenosis(MS), masked mitral regurgitation(MR) and dilated mitral valve annulus.

CASE REPORT

A 30 years old female presented with -

- progressive breathlessness over last 6 weeks
- Palpitation for 6 weeks
- Cough 6 weeks
- Not able to lie down in bed (orthopnea) 2 days

No history of-

- Chest pain
- Syncope
- Fever

She is not diabetic, hypertensive.

Clinical exam-

- Pulse- 115/min regular, equal on both sides.
- RR- 32/min
- Raised Jugular venous pressure
- Bilateral mild pedal oedema
- Mild hepatomegaly
- Bilateral basal rals

Cardiovascular system examination-

- Normally located apex impulse
- Leaft parasternal heave
- Loud pulmonary component of 2nd heart sound with wide split
- Normal intensity 1st heart sound
- Grade III/VI pansystolic murmur conducted to axilla
- Grade II/IV diastolic murmur in apical area

Cilinically she was suspected to have valvular heart disease and investigated with-

- Hematological and biochemical investigations were normal
- E.C.G- sinus tachycardia (rate 115 bpm), P-Pulmonale indicating right atrial hypertrophy
- Chest X-Ray- cardiomegaly of right ventricular type, prominent pulmonary conus.
- 2D Echocardiography/Doppler study-
- Large 3.5 × 3.1 mobile mass lesion attachecd to inter atrial septum(IAS) prolapsing in left ventricle during diastole obstructing mitral blood flow.
- Normal size left ventricle(LV) with mild systolic dysfunction LVEF 55%
- Dilated LA,LV, right atrium(RA)
- Moderate mitral regurgitation, severe tricuspid regurgitation
- Severe pulmonary hypertension, estimated pulmonary hypertension 62 mm of hg.
- Cardiac Magnetic Resonance Imaging(CMR)
- Non-dialated LV with mild systolic dysfunction, LVEF of 51%
- Dilated RV with systolic dysfunction, RVEF of 32%
- Mobile, large, pedunculated mass lesion in LA attached to inferior IAS with heterogenous enhancement, likely representing myxoma.
- Moderate MR with LA dilation
- No myocardial infarction, infiltration or inflammation

After stabilizing her hemodynamically she was referred for surgical removal of left atrial myxoma. Intra-operatively large friable left intra atrial myxoma around 3.5 x 3.2 was noticed and excised, mitral valve ring was dialated and coaptation of mitral valve leaflets was not proper resulting in significant mitral regurgitation. It was decided to repair mitral valve ring and put prosthatic valve.

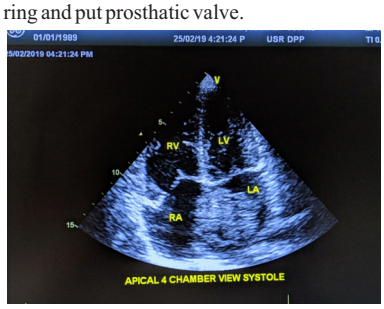

Fig.1 Apical 4 chamber view on 2D Echocardiography showing large myxoma in left atrium

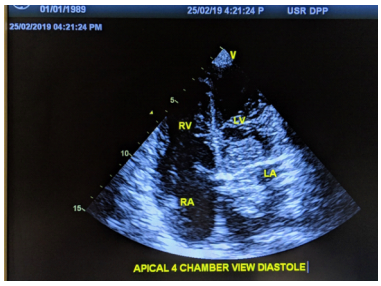

Fig.2 Apical 4 chamber view on 2D Echocardiography LA myxoma prolapsing in LV during diastole.

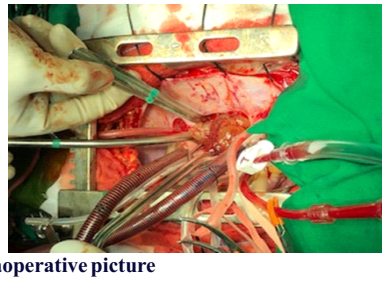

Fig.3. Intraoperative picture

DISCUSSION

Myxomas are primary benign tumors of heart most common in left atrium originating from inter-atrial septum(1) .In our case left atrial myxoma was originating from inferior IAS. They are asymptomatic as their development is slow , our patient was asymptomatic for long time. Embolism and intra-cardiac obstruction are common complications of atrial myxomas(1), however structural compromise giving rise to severe mitral stenosis is rare(2,5), our patient has features of functional mitral stenosis, with peak mitral valve flow gradient of 34.74 mm hg. The obstruction across mitral valve during diastole results in important hemodynamic consequence. A myxoma disturbs left ventricular inflow resulting in raised LA pressure and pulmonary hypertension. Our patient has features of severe pulmonary hypertension with dialated right atrium and ventricle with Right. Ventricular systolic dysfunction. The goal to achive hemodynamic stability is similar to mitral stenosis management, by maintaining myocardial contractibility, preloads, afterloads and keeping stable heart rate.

Mitral valve regurgitation in LA myxoma is due to annular dialation of mitral valve ring, improper coaptation of leaflets, direct damage to valve and/or subvalvular apparatus as myxoma body traverses through mitral valve during entire cardiac cycle. Prolapse of myxoma from LA to LV during each diastole lead to verying degree of mitral regurgitation causing volume overload to both LA and LV. The extent of mitral flow obstruction varies with body position as myxoma body itself interfere trans-mitral blood flow and also tends to mask mild, moderate and severe regurgitation. The continuous "wrecking ball" effect of the myxoma during each cardiac cycle against mitral valve apparatus in toto give rise to regurgitation and its severity is mainly dependent on resultant effect of myxoma body itself on the mitral valve(6-8). The grade and severity of regurgitation is mainly determined by body size of myxoma, length of stalk and to some extent verying degree of body position resulting in changes of trans-mitral blood flow (9). On routine 2D echocardiography the severity of regurgitation may be masked, similarly seen in our case both echocardiography and cardiac MRI showed moderate mitral regurgitation but intra-operative finding of mitral valve ring annular dialatation and improper cooptation of leaflets might have lead severe regurgitation. This implies that severity of regurgitation is not reliable parameter in myxoma for patients hemodynamic management and setting up proper surgical plan. In our case decision of annuloplasty and replacement of prosthatic valve was taken on table. In our day today practice we should have high index of clinical suspicion to detect intra-cardiac pathologies that could be missed with 2D echocardiography and cardiac MRI.

CONCLSION

Myxomas are primary benign tumors of heart commonest is LA myxoma. As they develop slowly they are asymptomatic, rarely due to mass effect they produce structural intra-cardiac pathologies. There can be functional mitral stenosis and most importantly mitral regurgitation may be masked, these flow abnormalities along with annular dilatation of mitral valve ring and improper cooptation of mitral valve leaflet can be missed on 2D echocardiography and cardiac MRI. So we should have high index of clinical suspicion to detect intra-cardiac pathologies in LA myxoma case. Keeping in mind structural pathologies due to mass effect of myxoma will help to set up proper surgical planning.

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