Case report

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Right Atrial Rhabdomyoma with Isolated Severe Tricuspid Valve Stenosis in an Adult: A Case Report

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Abstract

Cardiac rhabdomyoma in an adult without tuberous sclerosis complex is extremely rare entity.[8] Isolated tricuspid stenosis accounts for about 2.4% of all cases of organic tricuspid valve disease. A young woman presented to us with breathlessness and features of heart failure with a history of rheumatic fever. Eventually she was diagnosed with an atrial mass and an isolated tricuspid stenosis. Mass was excised and valve stenosis was corrected surgically under cardiopulmonary bypass. Histology proved the mass to be a rhabdomyoma. The patient is doing well till date.

Keywords: Adult; Heart rhabdomyoma; Tricuspid valve stenosis.

INTRODUCTION

Primary cardiac tumors are extremely rare entities, with a reported prevalence of between 0.0017% and 0.028%.[3] Rhabdomyoma is a rare benign tumor of mesenchymal origin and usually affecting the head, neck and limbs.[1] Cardiac rhabdomyomas are the most common cardiac tumors found in children (45%)[2] and usually associated with tuberous sclerosis. Adult cariac rhabdomyoma without

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tuberous sclerosis is not included in any literature till date.[8]

Tricuspid stenosis is a very rare valvular abnormality resulting from narrowing of the tricuspid valve, commonly found with other valvular pathologies, especially in rheumatic heart disease, but isolated disease is rare.[4] Tricuspid stenosis accounts for about 2.4% of all cases of organic tricuspid valve disease and is mostly seen in young women.[5,6] In a study tricuspid stenosis was found in 0.3% and pulmonary stenosis in 0.04% of all heart valves[7] pathologies.

CASE REPORT

A 37 year old female presented with chest pain, palpitation and breathlessness. she was suffering with such acute distress for past 2 weeks. She gave the history of occasional chest pain and breathlessness on exertion for past 1 year. She had a history of rheumatic fever and was on penicillin prophylaxis. She was married with two children. There was no history of any heart disease in her family. Her physical examination showed bilateral lower limb oedema, raised jugular venous pressure and mild jaundice with palpable, soft, non tender liver. No palpable lymph nodes were present. On auscultation, chest was clear with normal heart sounds without any added sounds. There was sinus tachycardia with features of right atrial enlargement, normal blood pressure. On blood examination, her liver enzymes were deranged with normal kidney function. Chest X-ray showed enlarged cardiac silhouette. On contrast enhanced computed tomogram a large right atrial hypo dense mass was seen (Fig. 1) which was supported by transthoracic echocardiography.

After the initial medical management patient was taken up for surgery. Under general anaesthesia, intra operative TEE (Transesophageal echocardiography) was done which showed a large right atrial mass with tricuspid valve flow acceleration (Fig. 2), signifying severe tricuspid valve stenosis. Other cardiac valves were normal.

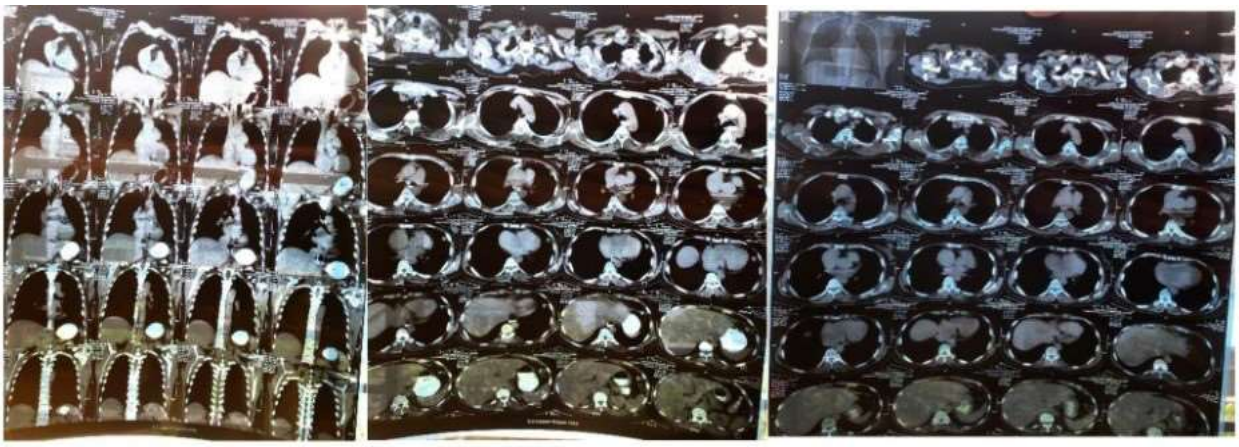

Fig. 1: Show the RA mass with enlarged right heart border

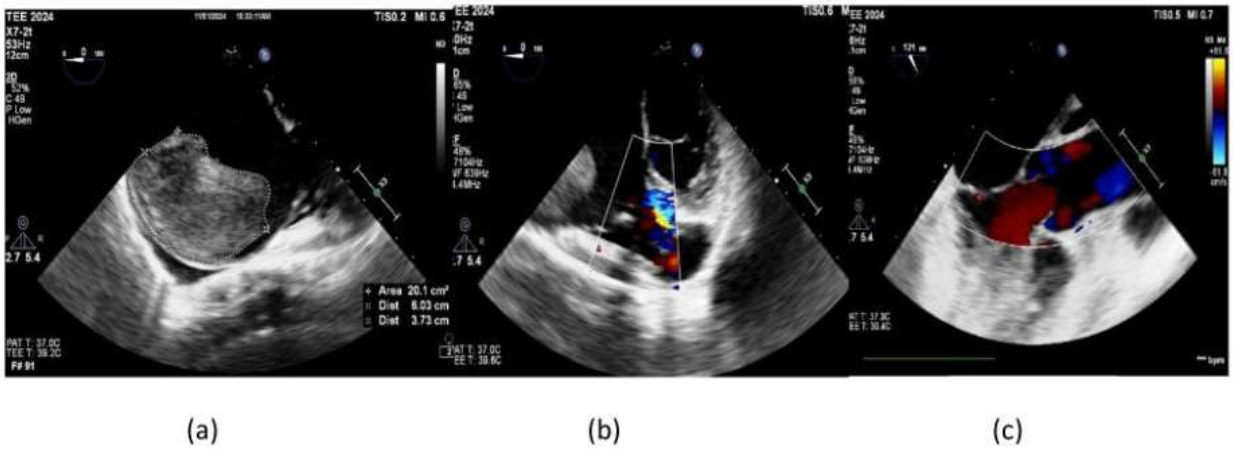

Fig. 2: (a) Right atrial mass (b) Tricuspid valve flow acceleration (c) Normal mitral and aortic valve

Operative management

Midline sternotomy and pericardiotomy were done and right atrium was seen to be grossly enlarged. After establishing cardiopulmonary bypass and arresting the heart, right atrium was opened and a firm mass of approximately 12×10 cm size was seen to be densely adherent with the wall of right atrium with atrial wall thickening. This mass was excised along with a portion of the right atrial wall. The tricuspid valve was examined which was fused, non-pliable with thickened leaflets. The valve opening was approximately 10mm size , just snugly fitting the head of cardiotomy suction (Fig. 3).

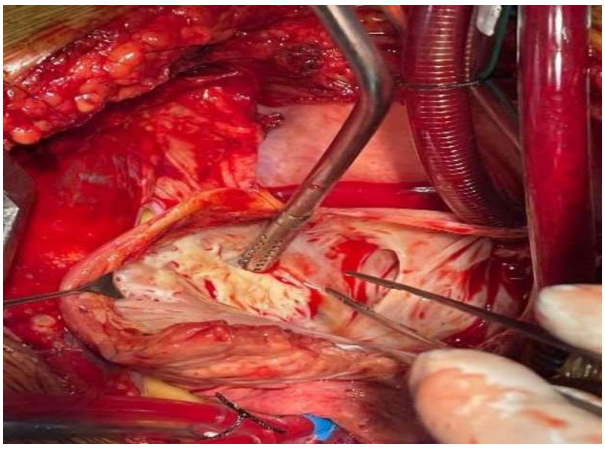

Fig. 3: The valve opening was approximately 10mm size, just snugly fitting the head of cardiotomy suction

Commissurotomy of leaflets were done and checked for any regurgitation by saline infusion test. Lastly, autologous pericardial (untreated) patch was used for reconstruction of the right atrium wall. Weaning from CPB was smooth and the post-operative period was also uneventful with minimum inotrope support. She was discharged in fifth post-operative day.

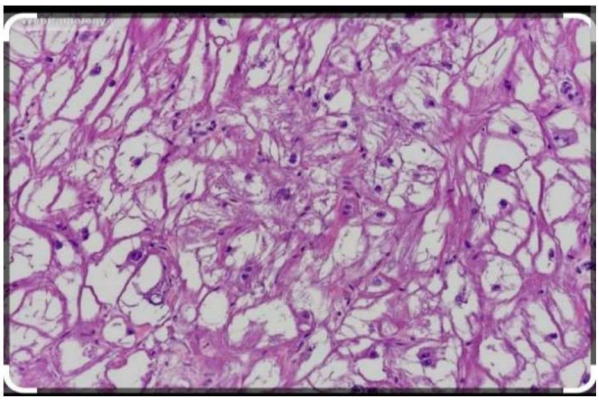

Fig. 4: Histology of the mass showing scattered cytoplasm and mixture of spindle cells and muscle fibers

Patient was followed up with histo-pathological report showed some scattered cytoplasm and mixture of spindle cells and muscle fibres, without any atypia suggesting pathological diagnosis of benign rhabdomyoma (Fig. 4)

DISCUSSION

Cardiac rhabdomyomas are rare in occurrence with generally benign in nature and of mesenchymal in origin.[1] This patient was presented with a large right atrial mass and isolated severe tricupid valve stenosis with 10mm opening and mean gadient around 9mmHg, causing excerberation of the symptoms and sought for urgent surgical intervention. Complete excision of the locally adhered tumor along with the wall of the right atrium is done, repair of the excised wall was done using autologus untreated pericardial patch and tricuspid stenosis was released with commissurotomy, Post operativly mild tricuspid regurgitation was there with vena contracta of 2mm.

CONCLUSION

Cardiac rhabdomyoma can be safely excised depending on the site and tissue involvement. Associated other pathology like congenital tricuspid stenosis could be repaired successfully with acceptable results.

REFERENCES

1. Hansen T, Katenkamp D. Rhabdomyoma of the head and neck: morphology and differential diagnosis. Virchows Arch. 2005;447:849-54.
2. Freedom RM, Lee KJ, Mac Donald C, Taylor G. Selected aspects of cardiac tumours in infancy and childhood. Pediatr Cardiol.2000;21:299-316.
3. Patel J, Sheppard MN. Pathological study of cardiac and pericardial tumours in a specialist UK Centre: surgical and autopsy series. Cardiovasc Pathol.2010;19:343-52.
4. Roberts WC, Ko JM. Some observations on mitral and aortic valve disease. Proc (BaylUniv Med Cent). 2008 Jul;21(3):282-99.
5. Tao G, Kotick JD, Lincoln J. Heart valve development, maintenance, and disease: the role of endothelial cells. Curr Top Dev Biol. 2012;100:203-32.

6. Roguin A, Rinkevich D, Milo S, Markiewicz W, Reisner SA. Long-term follow-up of patients with severe rheumatic tricuspid stenosis. Am Heart J. 1998 Jul;136(1):103-8.

7. Manjunath CN, Srinivas P, Ravindranath KS, Dhanalakshmi C. Incidence and patterns of valvular heart disease in a tertiary care high-volume cardiac center: a single center experience. Indian Heart J. 2014 May-Jun;66(3):320-6.

8. Ghosh S, Milunski MR. Cardiac Rhabdomyoma in Adult. Cureus. 2021 Apr; 13(4):e14565