Mystery of the Right Sided Heart Failure in a Young Patient

Gaurav Khandelwal*, Manish Ruhela, Rajeev Bagarhatta

Senior Resident, Department of Cardiology, SMS Medical College and Hospital, Jaipur, Rajasthan, India

*Corresponding author: gauravshri1231@yahoo.com

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Abstract  A 45 yr old female presented with clinical features of right side heart failure. 2D Echocardiography examination showed a homogenous, irregular, fimbriated mass attached to pulmonary valve, causing right ventricular outflow obstruction, with severe tricuspid regurgitation. To the best of our knowledge, this is the first case report of pulmonary valve mass, possible papillary fibroelastoma, which is large enough to cause right ventricular outflow obstruction, leading to severe tricuspid regurgitation and right sided heart failure.

Keywords: pulmonary valve, mass, right sided heart failure


1. Introduction

Pulmonary valve mass (fibroelastoma) is a rare cardiac tumour and it may cause right ventricular outflow obstruction, when sufficiently large in size. Many case reports in literature describe various masses on pulmonary valve, but this case report is unique because here we describe a large mass on pulmonary valve causing right sided heart failure due to right ventricular outflow obstruction, which was never described in the past.

2. Case Report

Figure 1. 2D Echocardiogram modified parasternal long axis view, showing mass attached to pulmonary valve causing right ventricular outflow obstruction
A 45 year old female presented with complaints of gradually progressive dyspnea on exertion for 6 months, abdominal distention and decreased appetite for 1 month. On examination, she had JVP up to 8 cm of blood column above angle of Louis, hepatomegaly up to 8 cm below sub costal margin, and free fluid in abdomen, single S2 (A2), ejection systolic murmur in left 2nd parasternal area and pansystolic murmur in left lower parasternal area. No other cardiac risk factors or significant past medical history were present. Haemogram and blood biochemistry were in normal limit. Ultrasound examination of abdomen showed congestive hepatomegaly. 2D Echocardiography showed a homogenous, irregular, fimbriated mass measuring 33 x 20 mm in RV outflow, attached to pulmonary valve, causing RV outflow obstruction (Figure 1). There was severe tricuspid regurgitation with RVSP = RAP + 54 mm Hg (Figure 2). RA and RV size were dilated and inferior vena cava (IVC) was dilated and noncollapsible. Patient was treated with diuretics and stabilized and was advised for further evaluation, but she refused. Patient was discharged on medical management.

3. Discussion

Differentials for the mass may be papillary fibroelastoma, cardiac myxoma, vegetation, nonbacterial thrombotic endocarditis (NBTE) or organized thrombus. Three out of four valvular tumors are papillary fibroelastomas. Papillary fibroelastomas are the second most common form of cardiac tumor, and more common in elderly. A valvular location is seen in 86% of cases – about 8 % are found on pulmonary valve [1,2]. The lesions are single in 91% of the cases, the average size is about 1 cm in diameter, and it can range from 0.2 to 4.6 cm. [1] The irregular, well demarcated, homogenous and fimbriated appearance of the mass on echocardiography in this case favours papillary fibroelastoma. Myxomas comprise 50% of all cardiac tumors. Myxomas mostly manifest in between the fourth and seventh decade, with female predominance. 83% of cardiac myxomas occur in the left atrium, occurrence in the ventricles is uncommon; only 0.6% of myxomas occur in the right ventricle.[3] Rarely, cardiac myxoma can involve heart valves directly[3.4]. Most of them have a smooth compact surface, and are pedunculated with a fibromuscular stalk. Cardiac myxoma is a less possibility in this case as the mass is irregular and fimbriated, it is directly attached to pulmonary valve; and pulmonary valve is very rare site for myxoma [5].

There was no predisposition for vegetation, like intravenous drug abuse or right sided catheterization in this case; there was also any history of fever. The pulmonary valve is least involved in patients with infective endocarditis and pulmonary regurgitation is often found. In NBTE, the vegetations are dense, smaller than 1.0 cm, broad based, and irregular in shape. NBTE has been reported in patients with advanced-stage malignancy, hematologic disorders, connective tissue disease, SLE, AIDS, hypercoagulable-state disorders & trauma from indwelling pulmonary or central venous catheter. No such condition was present in this patient.

There was no hypercoaguable condition or DVT in this patient, which could have lead to organized thrombus on the pulmonary valve.

The description of the mass and its location to ventricular aspect of pulmonary valve goes in favour of papillary fibroelastoma.

This is a rare case of a large mass on pulmonary valve, possibly papillary fibroelastoma causing RVOT.
obstruction leading to hypertensive severe tricuspid regurgitation.

To the best of our knowledge, about 50 cases of pulmonary valve papillary fibroelastoma have been described to date [1,2,6,7,8]. But most of them are small and do not disrupt the valve function, most of them have been found incidently at autopsy, and few as incidental finding in echocardiography examination [2,6].

4. Conclusion

Possibility of pulmonary valve mass should be considered in a patient with right side heart failure without any risk factors. This is probably the first case report of pulmonary valve mass described, possibly papillary fibroelastoma which is which is large enough to cause RVOT obstruction leading to hypertensive severe tricuspid regurgitation and right sided failure.

Statement of Competing Interests

Authors have no competing interests.

References