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Right Ventricular Outflow Tract Myxoma with Absent Pulmonary Valve

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ABSTRACT

Myxoma is the most common benign cardiac tumor. Absent pulmonary valves, often treated surgically in childhood, are associated with 3-6% of tetralogy of Fallot. It is unusual for absent pulmonary valves without other congenital heart diseases to be asymptomatic until adulthood. Here, we report the unique case of an 80-year-old female with right ventricular outflow tract myxoma and absent pulmonary valve that has, to our knowledge, never been reported. We successfully performed a simple mass resection and pulmonary valve implantation.

INTRODUCTION

Myxoma is the most common benign cardiac neoplasm with variable symptoms dependent on size and location. Around 75% of myxomas arise from multipotential mesenchymal cells in the left atrial oval fossa and often are accompanied by feeding vessels responsible for future recurrence after resection [Reynen 1995]. Here, we describe a rare case of right ventricular outflow tract (RVOT) myxoma with an absent pulmonary valve that has, to our knowledge, never been reported [Pemberton 2012; Mukadam 1994; Kasmeridis 2021].

Ethics statement: Institutional Review Board approval and clinical trial registration are not applicable for this study. The patient provided permission to publish case details, and patient identity has been protected.

CASE REPORT

An 80-year-old, otherwise healthy woman presented to our clinic with palpitations but no medical history of heart disease. On examination, a Levine 2/6 grade systolic murmur was noted in the pulmonary valve area, but laboratory data and chest X-rays were unremarkable. Transthoracic echocardiography found a mobile mass (23mm×16mm) attached to the infundibulum protruding from the RVOT into the pulmonary artery (PA). (Figure 1A) (Figure 1) PA peak velocity was mildly increased to 204 cm/sec, right ventricle pressure was estimated at 75 mmHg with mild right ventricle dilatation, and no significant pulmonary regurgitation (PR) was noted. There were no obvious abnormalities in the other heart valves and no congenital cardiac malformations, such as ventricular septal defects, were observed. Contrast-enhanced computer tomography showed a filling defect in the RVOT without peripheral PA embolic lesions (Figure 1B), while magnetic resonance imaging also revealed a mobile mass attached to the infundibulum beneath the pulmonary valve annulus, protruding into the PA. It was unclear if the pulmonary valve was intact or not (Figure 1C). But, since the mass had both a smooth rim and papillary-like structures, myxoma or papillary fibroelastoma was suspected and surgical intervention was planned.

A median sternotomy was performed. Under cardioplegic arrest, an incision was made from the anterior wall of the main PA to the RVOT upon which a smooth and jelly-like surface mass was confirmed beneath the intact pulmonary valve annulus; however, no pulmonary valve leaflets were grossly evident. The stalk of the mass was attached to the infundibulum, and the mass was completely resected (Figure 2A, 2B, 2C) before the attached site was cryo-ablated to prevent future recurrence. (Figure 2) Since there were no pulmonary valve leaflets, we performed pulmonary valve replacement using a 23mm Epic™ Supra Aortic Valve (Abbott, Inc. Abbott Park, IL). The anterior wall of the RVOT was reconstructed with a teardrop-shaped trimmed bovine patch. The posterior 2/3 circumference of the bioprosthetic valve cuff was sutured to the native pulmonary annulus and the anterior 1/3 circumference to the bovine patch. Cardiopulmonary bypass weaning was uneventful and transesophageal echocardiography confirmed good valvular function. Pathological findings were consistent with myxoma, and no valvular tissue was found (Figure 3A, 3B). (Figure 3) The patient discontinued mechanical ventilation on postoperative day 1 and was discharged home on postoperative day 19, with an uneventful postoperative course.

DISCUSSION

We experienced a unique case of RVOT myxoma with an absent pulmonary valve that has, to our knowledge, never been reported [Pemberton 2012; Mukadam 1994; Kasmeridis

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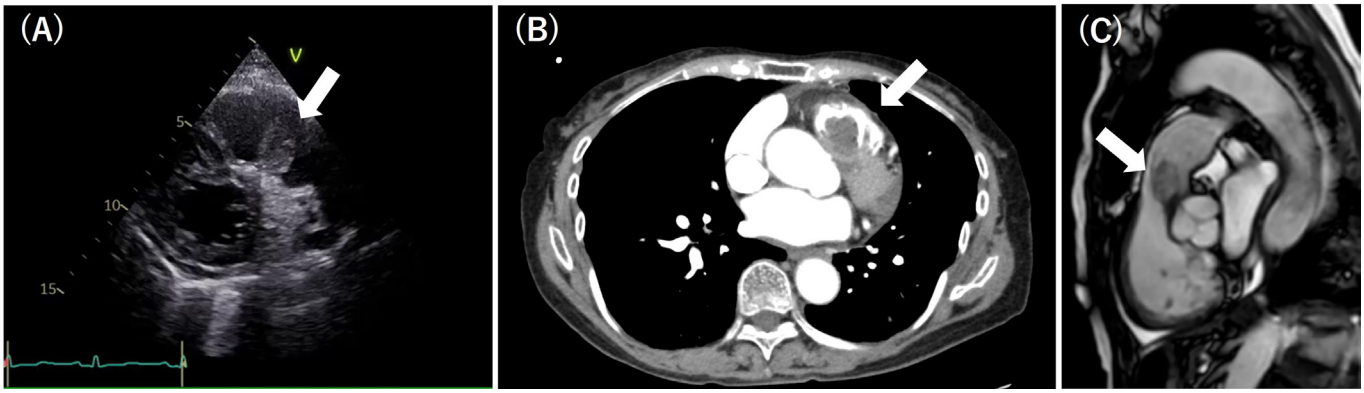

Figure 1. A) Transthoracic echocardiography examination reveals a mobile mass protruding into the pulmonary artery (white arrow); B) Computed tomography shows the filling defect in the right ventricular outflow tract (white arrow); C) Magnetic resonance imaging shows a mobile mass attached to the infundibulum beneath the pulmonary valve (white arrow).

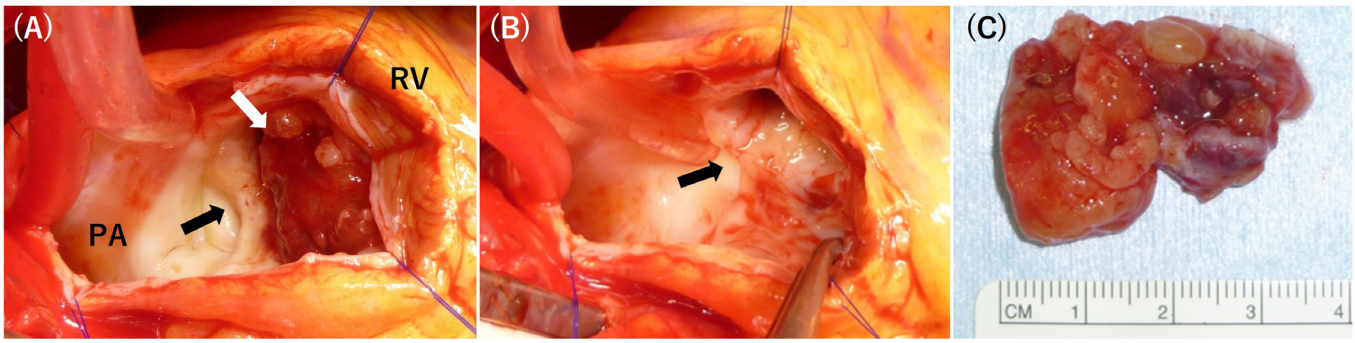

Figure 2. A) A visible mass underneath the pulmonary annulus (white arrow) and no pulmonary valve leaflet (black arrow); B) The pulmonary annulus was intact after mass resection (black arrow); C) The mass with a smooth, jelly-like surface. RV, right ventricle; PA, pulmonary artery.

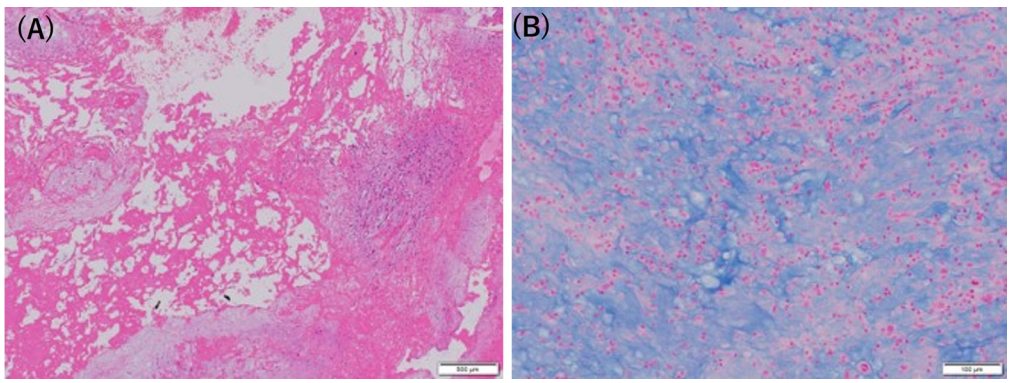

Figure 3. A) Typical pathology of myxoma without the valvular tissue (Hematoxylin and eosin stain, maginification×400); B) Periodic acid-Schiff stain with Alcian blue stain showing a basophilic mucinous matrix. (Magnification×200).

2021]. Although the valve leaflets were missing, the valvular annulus was present, so we considered the possibility of valve leaflet destruction due to malignancy or infective endocarditis. However, pathological findings showed a benign myxoma with no vegetation or other infectious findings around the RVOT. Therefore, we concluded that this case had a congenitally absent pulmonary valve and that the myxoma spontaneously developed. Absent pulmonary valves, often treated surgically in childhood, are associated with 3-6% of tetralogy of Fallot and 0.2-0.4% of total congenital heart disease cases [Allan 1994; Gottschalk 2017], while cases of isolated absent pulmonary valves are reported to be even rarer [Sayger 2000]. Tanabe et al. reported a case in which the patient was asymptomatic until age 59 [Tanabe 1992].

Our patient also had no cardiological symptoms until age 80. In this case, palpitations appeared at the age of 80, but

echocardiography showed only a mild increase in pulmonary artery flow velocity and mild right ventricular dilatation with no significant PR. Although the pathological development time is unknown, we posit that the RVOT became gradually stenotic, and pulmonary artery flow velocity increased due to the gradual growth of the tumor. As a result, it is possible that our patient, who had been asymptomatic for a long time, became aware of palpitations. In addition, since the tumor mimicked the function of the pulmonary valve, severe PR was not observed despite its absence. Generally, a simple resection is the primary surgical intervention for myxoma and provides excellent outcomes. However, in our case, postoperative severe PR was unavoidable with simple resection and therefore pulmonary valve replacement also was conducted. If the valves were partially preserved with mild pulmonary regurgitation expected, a simple tumor resection would have been acceptable.

In conclusion, we experienced a unique RVOT myxoma with absent pulmonary valve in which functional mimicry of the valve by the myxoma resulted in long-term symptom suppression. Simple resection of the tumor and pulmonary valve replacement resulted in an excellent outcome.

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