

MINI-FOCUS ISSUE: IMAGING

BEGINNER

IMAGING VIGNETTE: CLINICAL VIGNETTE

An Exceedingly Rare Case of Concomitant Quadricuspid Aortic Valve and Atrial Myxoma



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ABSTRACT

We present an exceptional case of a quadricuspid aortic valve associated with a left atrial myxoma. Both are rare conditions, and this association has not been reported yet. These conditions can be silent but may lead to several complications. This case highlights importance of a careful echocardiographic evaluation for early management. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2021;3:267-8) © 2021 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

A 57-year-old asymptomatic woman with a past medical history of aortic regurgitation was referred to our center (University Hospital of Grenoble, Grenoble, France) for investigation of an incidental atrial mass. Her blood pressure was 124/75 mm Hg, heart rate was 88 beats/min, and oxygen saturation while breathing room air was 98%. She reported neither dyspnea nor syncope. Auscultation revealed a 2/6 diastolic aortic murmur. Her lungs were clear. An electrocardiogram showed sinus rhythm with an incomplete left bundle branch block.

Transthoracic echocardiography showed a broad-based left atrial mass inserted on the atrial septum and moderate eccentric aortic regurgitation (**Figure 1A**).

A transesophageal echocardiogram confirmed a 27 × 19 mm heterogeneous mass, with areas of echolucency, attached to the left side of the interatrial septum (**Figure 1B, Video 1**), findings strongly suggestive of a myxoma. In addition, an unexpected quadricuspid aortic valve (**Figures 1C and 1D**) was found, with 2 equal larger cusps and 2 equal smaller cusps, corresponding to a type C according to Hurwitz and Roberts's classification (**1**). The coronary angiogram was normal.

Regarding the cardiac mass, a surgical procedure was chosen to prevent thromboembolic events. Aortic valve replacement with a no. 23 Carpentier-Perimount 3300 bioprosthesis (Edwards Lifesciences, Irvine, California) and concomitant complete tumor resection were performed. Post-operative follow-up was uneventful. Perioperative findings confirmed the quadricuspid nature of the aortic valve (**Figure 1E**). Histopathological analysis showed abundant mucoid matrix with perivascular arrangement and the presence of Gamna-Gandy bodies, thus confirming the diagnosis of myxoma (**Figure 1F**).

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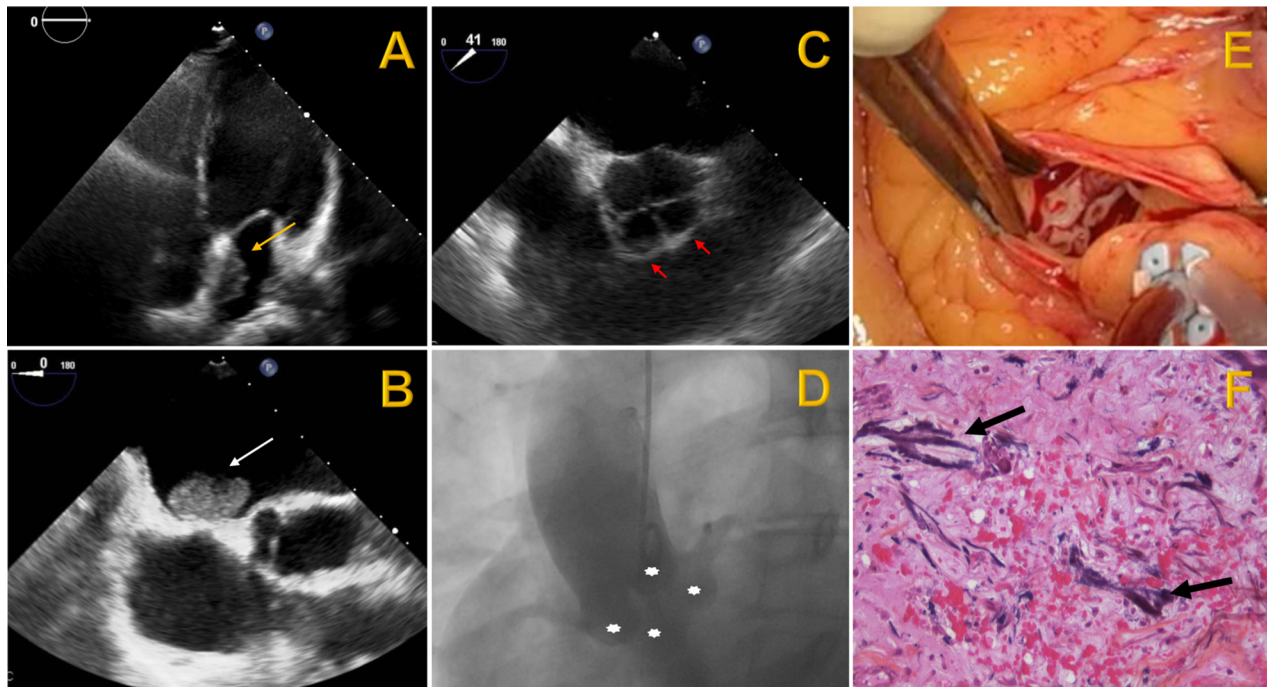
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This case highlights the coexistence of 2 exceedingly rare conditions: a quadricuspid aortic valve and an atrial myxoma. These latter may be silent but may lead to several complications, such as left ventricular systolic dysfunction and thromboembolic events (2,3).

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FIGURE 1 Quadricuspid Aortic Valve and Atrial Myxoma Assessment



(A) Transthoracic echocardiogram showing a left atrial mass with broad insertion (yellow arrow). (B) Transesophageal echocardiography showing a left atrial mass (white arrow). (C) Quadricuspid aortic valve shown by transesophageal echocardiography, with an accessory cusp (red arrows) in diastole. (D) Aortic angiography revealing 4 aortic valve cusps (asterisks). (E) Surgical excision of the myxoma. (F) Pathological results confirmed a left atrium myxoma with Gamna-Gandy bodies (black arrows). Hematoxylin and eosin stain, $\times 400$.

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KEY WORDS echocardiography, left atrial myxoma, quadricuspid aortic valve, valve replacement

APPENDIX For a supplemental video, please see the online version of this paper.