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Giant Right Atrium Syndrome

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Abstract

A case of rare giant right atrium in a woman with previous mitral replacement is described. The etiology is discussed in the light of literature reports.

Keywords: Atriomegaly; Tricuspid regurgitation; Rheumatic heart disease

Case Presentation

A 59-year-old woman presented to the clinic with a history of progressive dyspnea on exertion. Physical examination revealed jugular venous distension, an irregular pulse, and ankle edema, abdominal distension with liver enlargement 8-10 cm below the costal margin, free ascitic fluid and cachexia. The patient had undergone mitral valve replacement with a biological prosthesis in 1981 and subsequently with a mechanical prosthesis in 1996 for rheumatic mitral insufficiency, both times through a sternotomy approach. A chest radiograph showed near-complete opacification of the right upper-mid lung zones, due to marked bulging of the right heart border (Figure 1). A transthoracic echocardiography revealed severe tricuspid insufficiency, massive dilatation of the right atrium with a calculated volume of 700mL, mild dilatation of right ventricle with fractional area change of 44% and normal mitral prosthesis function. Left ventricular ejection fraction was normal. There was no evidence of organic lesions of the tricuspid valve. Cardiac catheterization showed normal coronary arteries and pulmonary pressures. CT scan demonstrated a huge right atrium, (Figure 2) chronic liver disease, moderate as cites and significantly increased diameter of the inferior vena cava. Intraoperative assessment confirmed the diagnostic findings (Figure 3). The patient underwent tricuspid valve replacement with a 31mm biological prosthesis (Figure 3). The post-operative course was uneventful and the patient was transferred to rehabilitation 10 days later.

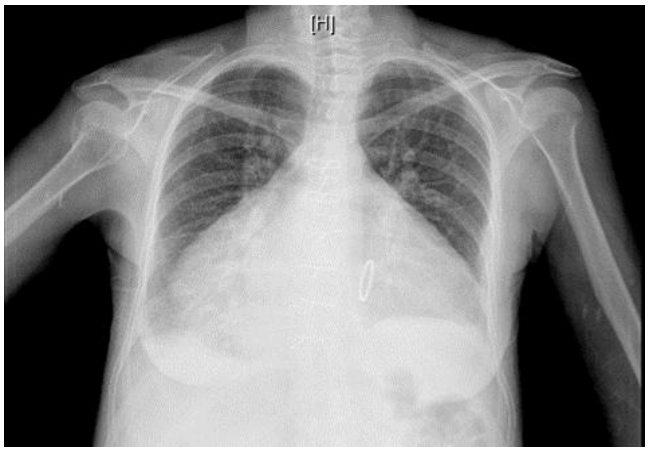

Figure 1: Chest radiograph showing biological valve prosthesis and markedly enlarged cardiac silhouette: antero-posterior view.

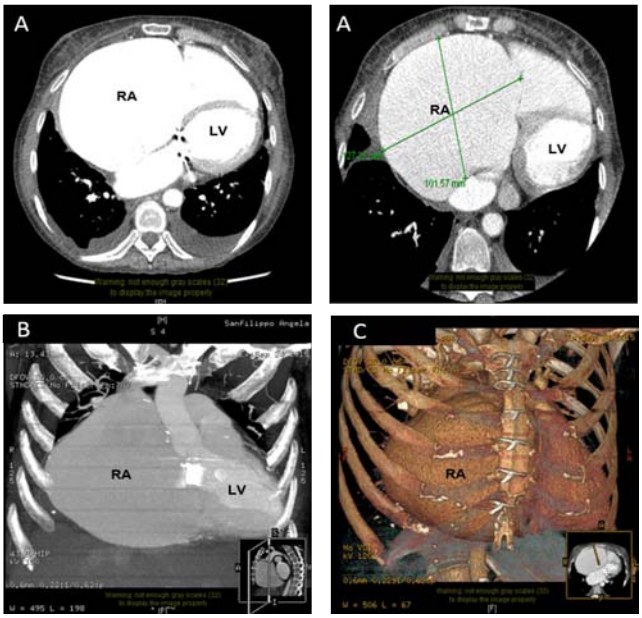

Figure 2: Chest computer tomography images showing giant right atrium and biological valve prosthesis. Axial (a), coronal (b) views and 3D CT reconstruction (c) RA, right atrium; LV, left ventricle.

Discussion

Giant left atrium was first described by Hewett in 1849 and represents a significant risk factor in patients undergoing mitral valve surgery [1]. On the other hand, we found only tree reports of a non-idiopathic giant right atrium in the adult [2-4]. The most common causes of an enlarged right atrium in adults are chronic pulmonary disease, severe mitral abnormalities with pulmonary hypertension, pulmonary emboli and tricuspid stenosis [2]. Malformations with massive enlargement of the right atrium and coronary sinus are rare and usually associated with congenital heart disease in infants and

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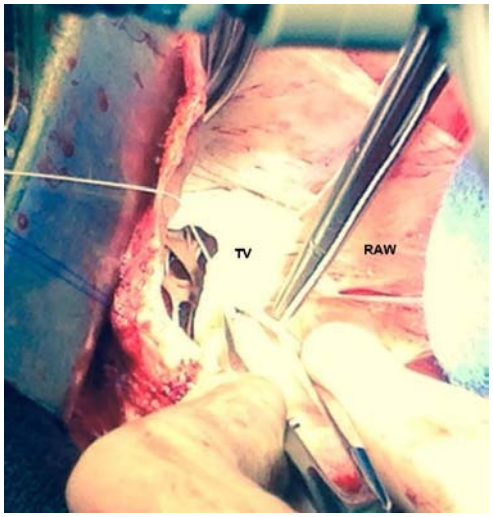

Figure 3: Surgical view: the huge right atrium and tricuspid valve. TV: Tricuspid Valve; RAW: Right Atrium Wall.

children although some idiopathic cases have been reported [5]. This case represents one of the largest giant right atrium described in adults. We believe that this patient has the extremely rare condition of severe functional enlargement of the right atrium leading to dilatation of tricuspid annulus, severe tricuspid regurgitation and atrial fibrillation with subsequent right heart failure.

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