Massively dilated right atrium masquerading as a mediastinal tumor

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Abstract

Severe tricuspid valve insufficiency causes right atrial dilatation, venous congestion, and reduced atrial contractility, and may eventually lead to right heart failure. We report a case of a patient with severe tricuspid valve insufficiency, right heart failure, and a massively dilated right atrium. The enormously dilated atrium compressed the right lung, resulting in a radiographic appearance of a mediastinal tumor. Tricuspid valve repair and reduction of the right atrium was performed. Follow up examination revealed improvement of liver function, reduced peripheral edema and improved New York Heart Association (NYHA) class. The reduction of the atrial size and repair of the tricuspid valve resulted in a restoration of the conduit and reservoir function of the right atrium. Given the chronicity of the disease process and the long-standing atrial fibrillation, there is no impact of this operation on right atrial contraction. In combination with the reconstruction of the tricuspid valve, the reduction atrioplasty will reduce the risk of thrombembolic events and preserve the right ventricular function.

Introduction

The functional role of the right atrium is comprised of three components: a reservoir function, a conduit function, and a pump function to complete right ventricular filling.¹ Muscular fibres and a trabecular network, as well as a normally functioning pericardium, are important to maintain the structural integrity of the right atrium. Previous studies have shown that the atrial reservoir function is four times more important than active atrial contraction in providing energy for the work of ventricular filling.²

Tumors of the mediastinum are uncommon lesions that may be discovered by chest x-ray. We herein report a patient with severe tricuspid insufficiency and secondary dilatation of the right atrium resulting in a radiographic appearance of a mediastinal tumor.

Case Report

A 68-year-old female presented to our hospital with dyspnea [New York Heart Association (NYHA) class IV], peripheral edema, and cirrhosis [gamma-glutamyltransferase (GGT) 13-times higher than normal] with evidence of hepatic congestion. The patient was known to have chronic pulmonary hypertension [33 mmHg + central venous pressure (CVP)], jugular venous distention, chronic atrial fibrillation, and a normochromic anaemia. The patient was treated in 1976 for rheumatic mitral stenosis, at which time an open mitral valvuloplasty was performed. In 1985 the patient developed recurrent symptomatic mitral valve stenosis in combination with aortic valve stenosis. Mechanical valve replacement of the aortic and the mitral valve (both tilting disc prosthesis) was performed at that time. Echocardiographic findings at the time of admission consisted of an enormously dilated right atrium (71 x 155 mm), a dilated left atrium (63 x 48 mm) and a moderately dilated right ventricle. The patient also had severe tricuspid valve insufficiency secondary to annular dilation (51 mm) with no visible coaptation between the leaflets. The function of the aortic and mitral mechanical prostheses was good, with a mildly elevated gradient across the mitral valve (Pmean 6 mmHg). Both ventricles showed no restricted myocardial contractility without hypokinetic or akinetic areas. However, due to the high-grade tricuspid valve insufficiency, an accurate assessment of the right ventricular ejection fraction could not be obtained. Chest X-ray suggested a right-sided mediastinal tumor with complete compression of the middle and lower lobes of the right lung (Figure 1). A computed tomography (CT) scan confirmed that the right atrium was responsible for compression of these structures (Figure 2, Figure 3). Because of the symptoms and findings, the decision was made to repair or replace the tricuspid valve and to reduce the size of the right atrium (reduction atrioplasty).

Although the combination of enlarged right atrium, atrial fibrillation and turbulent blood flow on account of the regurgitation represented a strong predisposition for the development of thrombus formation, both echocardiography and CT showed no atrial or ventricular thrombus. Intraoperatively, no thrombus was identified. The reason for the lack of thrombus may have been the long-standing therapy with Coumadin for the mechanical valves.

The right atrium was known to lie in direct contact with the posterior table of the sternum by CT scan, and therefore the decision was...
made to cannulate the right femoral and internal jugular veins for cardiopulmonary bypass (CPB). Arterial cannulation was performed via the right femoral artery. During full flow CPB, as much blood volume as possible was extracted from the patient’s body and ventilation was stopped while a median sternotomy was performed with an oscillating saw. With these manoeuvres, we managed to avoid injury to the cardiac structures. It was subsequently necessary to dissect away the right atrium from the lung, because of extensive adhesions between these two structures. Because of the previous two cardiac operations, no identifiable pericardium was found. The giant right atrium was opened and an extensive trabecular network, similar to the inside of the right ventricle, was observed (Figure 3). The leaflets of the tricuspid valve were normal in appearance, but the annulus was markedly dilated. A tricuspid valve repair was performed using a 30 mm Carpentier Edwards Classic ring (Figure 3; Edwards Lifesciences, Irvine, CA, USA). Injection of saline into the right ventricle opened and an extensive trabecular network, revealed a competent valve without any insufficiency. A reduction atrioplasty was subsequently performed using an extensive plication technique. To avoid new adhesions, the right atrium was covered anteriorly with Seprafilm (Genzyme, Konstanz, Germany) prior to closure of the sternum.

The postoperative course was complicated by the development of pneumonia in the newly ventilated parts of the right lung. Culture-guided antibiotic therapy resulted in convalescence of the pneumonia over the next few days. Three weeks after the operation, the patient was discharged from the hospital in good condition. Six weeks after discharge, the patient was examined in our out-patient clinic. Her dyspnea and exercise level were improved (NYHA II-III) and the peripheral edema had resolved. The liver function tests had also improved, with a GGT level 8 times normal. Postoperative echocardiographic findings consisted of mild central tricuspid regurgitation (vena contracta 2 mm) and a markedly reduced size of the right atrium (31×70 mm; Figure 1).

**Discussion**

Patients with severe tricuspid valve regurgitation are a poor prognostic population. Chronic volume overload gives rise to atrial dilatation and decreased contractility and compliance. The trabecular network of the right atrium expands during chronic volume overload and may result in an appearance that is similar to that found in the ventricle. Microstructural findings consist of large deposits of collagen fibres and perivascular fibrosis. The amount of β-myosin chains in the muscle increases. The observed atrial changes in the case reported herein are due to a combination of chronic volume overload, from the tricuspid valve insufficiency, as well as chronic pressure overload, from the chronic pulmonary hypertension. Obviously, a regular contractile function of the right atrium will never be restored. The reasons for this conclusion are the preoperative course of the patient with long-standing atrial fibrillation, massive atrial dilation and the age of the patient. Additionally, the reduction atrioplasty prevents the development of an effective contractile power. Because of this expectation, we strongly recommend therapeutic anticoagulation with Coumadin to prevent the development of thrombus.

**Conclusions**

The reduction of the atrial size and repair of the tricuspid valve resulted in a restoration of the conduit and reservoir function of the right atrium. Given the chronicity of the disease process and the long-standing atrial fibrillation, there is no impact of this operation on right atrial contraction. In combination with the reconstruction of the tricuspid valve, the reduction atrioplasty will reduce the risk for thrombembolic events and preserve the right ventricular function.

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