Multiple Fibroelastomas in a Patient With Complex Congenital Heart Disease
Complications and Outcome

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A 42-year-old man with a previous diagnosis of congenitally corrected transposition of the great arteries, a large ventricular septal defect (VSD), Ebstein anomaly of the tricuspid valve, and mild pulmonary stenosis was referred after large “vegetations inside heart” had been demonstrated at routine follow-up echocardiographic examination. Four years before, he began having episodes of atrial fibrillation and bradycardia. DC cardioversion had failed, and a permanent pacemaker was implanted 2 years later. There was no history of febrile illness or history to suggest systemic embolization, and warfarin had always been well maintained at therapeutic levels. On examination, he looked well and pink with no clubbing. On auscultation, he had a soft early ejection systolic murmur at the left sternal edge and a loud single second heart sound. There were no stigmata of infective endocarditis and no splenomegaly. Echocardiography showed several large mobile echogenic masses attached to the abnormal tricuspid valve, VSD border, and pulmonary valve (Figure 1). The systemic right ventricle was small, with severely impaired ventricular function. The subpulmonary left ventricle was mildly dilated with moderately impaired function. There was mild pulmonary regurgitation with an estimated end-diastolic pulmonary artery pressure of 60 to 70 mm Hg. Repeated blood cultures and all inflammatory markers were negative. He had significant pulmonary hypertension and was therefore unsuitable for surgery.

One year later, he had an attack of syncope that was assumed to be embolic. Amiodarone was started together with antifailure treatment because his ventricular function was deteriorating. During the next 2 years of follow-up, he had a stroke resulting in right hemiparesis and severe expressive dysphasia and multiple pulmonary emboli, which was complicated by infection. He died of cardiopulmonary failure 4½ years after initial finding of intracardiac masses.

At postmortem examination, the heart was enlarged and the anatomic findings were consistent with congenitally corrected transposition of the great arteries, a large VSD, Ebstein anomaly of the tricuspid valve, and mild pulmonary valve stenosis. There were multiple fingerlike masses

Figure 1. A, Transthoracic echocardiogram of the apical 4-chamber view showing atrioventricular discordance with abnormal left-sided tricuspid valve (Ebstein anomaly) and a large VSD. Multiple echogenic masses (arrows) are attached to the tricuspid valve and ventricular septum. B, Apical 5-chamber view showing the pulmonary artery (with bifurcation) arising from the right-sided morphological left ventricle; the pulmonary valve is thickened and doming, with a small echogenic mass attached (arrows).
inside the cardiac chamber close to the VSD on both sides and extending to the left-sided tricuspid valve (Figure 2). These were variable in size, with an average diameter of 1.5 cm. Similar masses were attached to the pulmonary valve leaflets. Histology confirmed that they were fibroelastomas (Figure 3).

To the best of our knowledge, multiple fibroelastomas in a patient with congenitally corrected transposition of the great arteries have not been reported previously.

**Disclosures**

None.
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