Case 4/2012 – 25-year-old man with obstructive fibroma in right ventricular outflow tract

Edmar Atik

Instituto do Coração do Hospital das Clínicas da Faculdade de Medicina da Universidade de São Paulo, São Paulo, SP - Brazil

Clinical data: During routine examination, cardiac murmur was heard in an 8-year-old patient. At the time, infundibular pulmonary stenosis with pressure gradient of 69 mmHg was diagnosed by echocardiography. Progressive fatigue on efforts has been noticed for two years, as well as unstable chest pain. No specific medication was administered during this period.

Physical examination: The patient was in good general condition, eupneic, with normal pulse and normal color. Weight: 42.6 kg; height: 160 cm; BP: 110/70 mmHg; HR: 80 bpm, O2 saturation: 96%. The aorta could not be felt at suprasternal notch.

Precordial systolic impulsions were discrete. Apical impulse was not palpable. The heart sounds were normal and systolic murmur ++ was harsh, across the left sternal border. The liver was not palpable and the lungs were clear.

Complementary tests:

Electrocardiogram showed sinus rhythm with signs of severe right ventricular overload with Rs morphology and negative T wave in V1, AP: +60º, AQRS: +110º, AT: +60º (Fig. 1).

Chest radiography shows cardiac area within normal limits, with rounded morphology. The middle arch is rectified and the pulmonary vasculature is slightly increased (Fig. 1).

Echocardiography (Fig. 2) showed hypertrophy of the right ventricular wall. There was a mass in the ventricular septum protruding into the right ventricular outflow tract causing a pressure gradient of 70 mmHg. The pulmonary valve was preserved, as well as biventricular function. The measures corresponded to RV =32 and LV = 34.

Nuclear magnetic resonance imaging showed the same aspects with marked obstruction in the right ventricular outflow tract (Fig. 2).

Cardiac catheterization also showed obstruction in the right ventricular outflow tract and pressure values were RV = 80/13, right ventricular outflow tract = 27/8, PT 27/15-19, LV = 100/14 mmHg.

Keywords

Ventricular septum; heart neoplasms; ventricular dysfunction, right, cardiac fibroma

Mailing Address: Edmar Atik •
InCor - Av. Dr. Enéas Carvalho de Aguiar, 44 - 05403-000 - São Paulo, SP - Brazil
E-mail: conatik@incor.usp.br

Clinical Diagnosis: Right ventricular outflow tract stenosis by protrusion of tumor mass in the interventricular septum by myocardial fibroma

Clinical reasoning: Clinical findings were consistent with the diagnosis of congenital heart disease accompanied by long-lasting obstruction of the pulmonary flow. Radiographic elements, such as increased pulmonary blood flow and middle arch rectified also lead to the diagnosis of pulmonary stenosis. Other imaging elements were decisive for the diagnosis of the cause of pulmonary stenosis, dependent on tumor mass protrusion in the right ventricular outflow tract from the interventricular septum. Given the great impact of this longstanding obstruction, with limiting symptoms and diagnosis of tumor mass in the ventricular septum with greater protrusion into the right ventricle, a presumptive diagnosis of fibroma was established.

Differential diagnosis: Other heart diseases associated with pulmonary stenosis can also appear in a similar way in clinical terms, such as double right ventricular outflow tract, double inlet single ventricle and other similar functional anomalies.

Conduct: By median thoracotomy, the ventricle was resected by partial longitudinal right ventriculotomy, whitish and hard septal mass obstructing the outflow of this ventricle, suggestive of myocardial fibroma. For fear of damaging the electrical conduction system and iatrogenic ventricular septal defect, it was allowed to leave a residual tumor in the region that caused further pressure gradient of 50 mmHg. In the immediate postoperative period, performed without complications, harsh systolic murmur of ++ intensity, was still listened, despite a feeling of easier breathing reported by the patient.

Comments: The diagnosis of obstruction of right ventricular outflow tract was not difficult given the presence of typical clinical elements and images. The cause of the stenosis due to tumor mass was guided by echocardiography, which, due to exacerbated echogenecity distinct from the rest of the septum and ventricular wall, referred to the possibility of myocardial fibroma. This hypothesis was confirmed by direct operative view and anatomopathological study. This tumor rarely develops similarly to the usual congenital pulmonary stenosis. This simulation resulted from the restriction of the tumor to a certain portion of the interventricular septum. Usually, the fibroma is more invasive in the myocardium, causing myocardial dysfunction and arrhythmias, which are causes of unfavorable outcomes, at an earlier age, in children and infants.

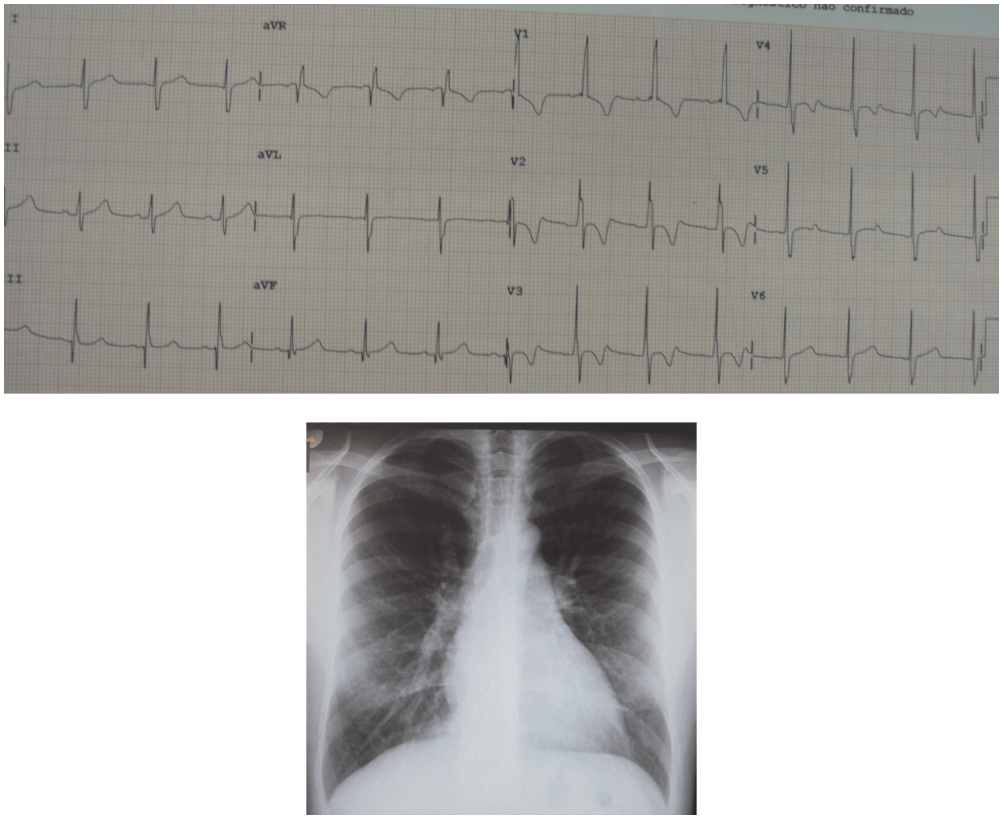

Figure 1 - *Chest X-ray reveals normal cardiac area with rectified medium arch and pulmonary vasculature increased in the pulmonary hila. This image is compatible with heart diseases that accompany long-standing pulmonary stenosis. Electrocardiogram reveals the signs of sharp right ventricular overload.*

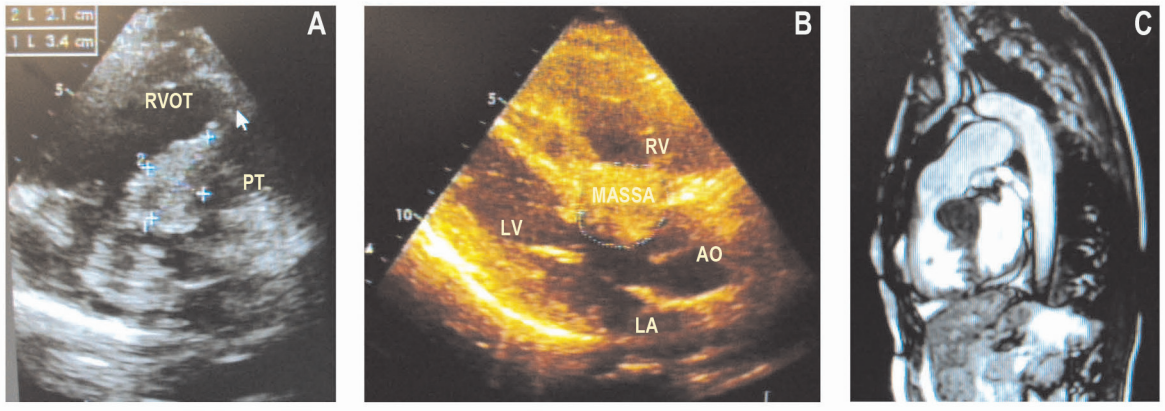

Figure 2 - *Echocardiogram shows an obstructive mass in the right ventricular outflow tract in parasternal short axis at A, restricted to the interventricular septum in longitudinal section in B, and clear obstruction of right ventricular outflow tract on magnetic resonance imaging in C.*