Fibroma of the Left Ventricle in a Patient with Sotos Syndrome

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Primary tumors of the heart are rare, with an incidence between 0.001% and 0.33% in autopsy findings, and with fibromas representing about 4% of benign cardiac tumors. We report the case of a cardiac fibroma in a 5-year-old child affected by Sotos syndrome. The mean sign was a ventricular tachycardia. (ECHOCARDIOGRAPHY, Volume 18, February 2001)

Sotos syndrome, cardiac fibroma, cardiac tumors, ventricular tachycardia, echocardiography

Sotos syndrome was described for the first time in 1964.1 It is characterized by macrodolichocephaly with a prominent and rounded forehead, accentuated by frontoparietal balding and hypertelorism. Other common features of Sotos syndrome are large size at birth, excessive growth during childhood, delay in motor and speech development, and awkward gait.1-4 Its etiology is unknown but autosomal-dominant or autosomal-recessive mode of inheritance has been suspected.3 It seems, however, that there are no chromosomal abnormalities. To date, the literature has reported on approximately 300 cases of Sotos syndrome. According to the literature, individuals with Sotos syndrome have a higher risk for tumors than those without the syndrome.5,6 Indeed, Sotos patients have a higher incidence of many tumors, such as Wilms’ tumor, neuroblastoma, liver, and lung carcinoma. The reason for the high incidence of neoplasms is not yet clear.5,6 A cardiac fibroma was found in a patient with Sotos syndrome observed by us.

Case Report

A 5-year-old boy, affected by Sotos syndrome, was referred for evaluation after a syncope. At the age of 3, the patient was diagnosed with acute lymphoblastic leukemia, successfully treated with complete remission. On physical examination there was no murmur but pulse was irregular. Electrocardiogram (ECG) showed a sinus rhythm frequently interrupted by ventricular premature beats and inverted T waves across the chest leads. Furthermore, several episodes of sustained ventricular tachycardia were recorded in 24-hour continuous ECG.

Two-dimensional echocardiography showed an echo dense, oval-shaped mass 2x3 cm in diameter located within the posterior wall of the left ventricle. The mass extended itself from the base to the mid-ventricle and was quite homogeneous. It infiltrated the myocardium and protruded into the ventricular cavity (Fig. 1).

The left ventricle size was normal but systolic function was quite reduced (ejection fraction = 50%). Cardiac valves, atria, pericardium, and right ventricle were normal. A thoracotomy was performed but unfortunately the tumor could not be removed because it extensively infiltrated the posterior wall of the left ventricle. However, histology proved the tumor to be a fibroma. The possibility of a heart transplant was discarded because the risk of a recurrence of the patient’s leukemia was considered high.

After the patient had other symptomatic episodes of sustained ventricular tachycardia, a therapeutic attempt was performed with Propafenone, Xilocaine, and Propanolol. As they were ineffective in suppressing the ventricular tachycardia, Amiodarone (200 mg/day P.O.) was administered and proved effective on arrhythmias, as shown by the lack of changes after programmed ventricular stimulation of the right ventricle.
The patient has remained well through a 5-year follow-up. No life-threatening arrhythmias at the 24-hour ECG monitoring and no undesired side effects to the drug have been recorded.

Discussion

A comprehensive review of the literature indicates that Sotos syndrome has never been reported in association with a cardiac fibroma. Cardiac fibroma is a benign tumor that accounts for about 5% of primary cardiac tumors. It can develop at any age, both in males and females. However, it is more frequently observed in children and one-third of the cases are diagnosed in infants of less than 1 year. Actually, fibroma represents the second most common tumor in infancy after rhabdomyomas. Unlike rhabdomyoma, cardiac fibroma is usually single, noncapsulated, and not yet invasive, and has a diameter ranging from 3–8 cm. As a rule, it is intramural and involves the left ventricle but in rare cases invades atria and the right ventricle.

Histologically, cardiac fibroma is formed by collagen fibers and fibroblasts. It frequently is calcified in the central part and at times appears cystic. Its clinical manifestations are protean and mainly depend on size and location. Its most undesirable trait is obstruction of the outflow tracts since this can determine heart failure.

Arrhythmias occur frequently and often are manifested by dizziness and palpitations. Sometimes they can be life threatening, either in the form of a complete atrioventricular block or a ventricular tachycardia. When the mass is localized near the conduction system, syncope or sudden death can occur. ECG usually reveals abnormalities, including bundle branch block and ventricular hypertrophy; ST-T wave abnormalities are invariably present.

On echocardiography, cardiac fibroma appears as a single and bright mass within the left ventricular wall or the interventricular septum. Due to its location, it is often unsuitable for surgical removal. At any rate, surgery should be performed when there is an inflow-outflow obstruction causing cardiac failure.

Our case is of some interest because of the association of a fibroma with Sotos syndrome and because of the previous acute lymphoblastic leukemia. We believe that both ECG and two-dimensional echocardiography should be performed in all patients with Sotos syndrome in order to rule out cardiac tumors and to prevent sudden deaths.

References

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LV FIBROMA IN SOTOS SYNDROME


