CASE REPORT

PEDIATRIC ATYPICAL SHONE'S COMPLEX

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Abstract

Shone's complex is an extremely rare and severe congenital heart disease with about fifty cases reported in literature. It is characterized by great clinical variability and difficulties in diagnosis and management. Shone's complex consists on the association of obstructive defects at many levels of the left ventricular outflow tract. The full or typical Shone's complex, in its first description, corresponds to 4 levels of obstruction - supravalvular mitral membrane, valvular mitral stenosis due to parachute mitral valve, subaortic stenosis, and coarctation of the aorta. Since then, several Shone's complex descriptions with three or two levels of obstruction have been reported and called incomplete Shone's complex or atypical. We report the case of a 57-day-old boy transferred to pediatric department for persistent dyspnea. The diagnosis of atypical Shone's complex with three levels of obstruction was made. Surgical treatment was performed at 60th day of life with favorable outcome.

Keywords : Shone's complex, acute respiratory distress syndrome, diagnosis

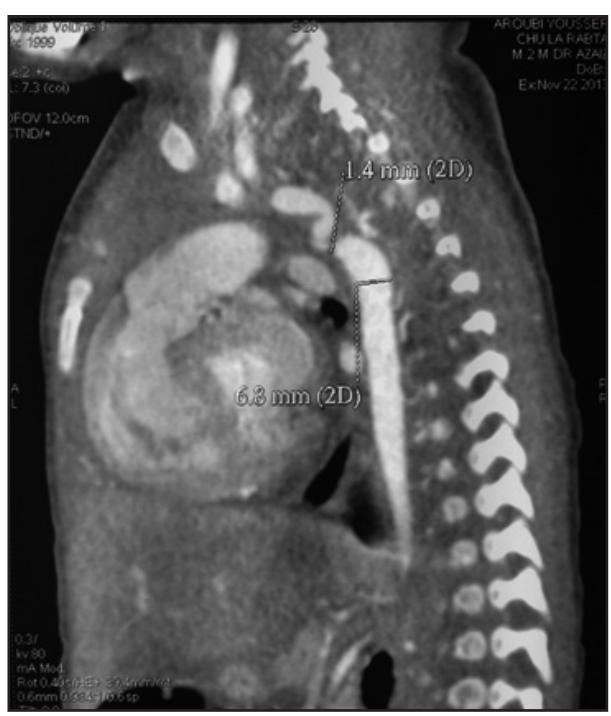
Introduction

Shone's complex is a rare condition, first described by Shone in 1963. (1) It consists of multiple left ventricular outflow tract obstruction. The components include supravalvular mitral membrane, valvular mitral stenosis due to parachute mitral valve, subaortic stenosis, and coarctation of the aorta. In clinical practice, the definition of Shone's anomaly has been extended beyond the original Shone complex, to include patients with additional forms of left heart anomalies. We report the case of an infant presenting a severe dyspnea. Echocardiography and computed tomography angiography revealed atypical Shone's complex.

Case Report

A 57-day-old male infant was referred to our department for management of dyspnea. He was born by normal vaginal delivery to parents of first degree consanguineous marriage. The pregnancy was uneventful and prenatal ultrasounds were normal. Mother was a 32-year-old primigravida. Apgar scores were of 9 and 10 at the 1st and 5th minutes, respectively. His birth weight was 3150g (75th percentile). He had a normal physical examination at birth. At 55th day of life, the baby presented with dyspnea with feeding difficulties and was admitted to our department. On examination, weight was 3770g, he was hypothermic, had respiratory distress with tachypnea and retraction signs. The extremities were cold. He was cyanotic with oxygen saturation at 85% in the air. Cardiac auscultation revealed a gallop and single 2nd heart sound. There was hepatomegaly. Femoral pulses were not found though axillary pulses were present suggesting aortic coarctation. Blood pressure was 125/80 mm of Hg in the upper extremities and 90/50 in the legs. On chest X-ray there was cardiomegaly, hepatomegaly and increased pulmonary vascular markings. Electrocardiogram (ECG) showed left ventricular hypertrophy. 2-D transthoracic echocardiogram (2D Echo) showed tightly bound isthmic coarctation of the aorta with a small aortic arch and a pre structural patent ductus arteriosus shunting exclusively the aorta and pulmonary artery with a flow of 4m s-1. There was a severe congenital mitral stenosis with the gradient between the left auricle and the left ventricle at 8 mmHg. Aortic valve was bicuspid with aortic stenosis. There was a left ventricular hypertrophy. The maximum gradient between the left ventricle and the aorta was estimated to be 19 mmHg. This gradient was underestimated because of the coarctation of the aorta. The pulmonary arterial pressures were normal. Computed tomography angiography showed aortic coarctation and left ventricle hypertrophy (Figure 1). He was treated with oxygen, furosemide, alprostadil and surgical coarctation repair was performed on the 60th day of life. Correction of mitral stenosis will be planned for the future. The postoperative course was uneventful.

Figure 1: Computed tomography angiography showing aortic coarctation



Discussion

In the complete form of Shone complex, all the four obstructive lesions on the left side of the heart are present. However, the incomplete forms, called also atypical or form fruste, consists only of two or three lesions. The rate of obstruction on 4 levels is 7.9%, three levels 26.3% and two levels 65.8%. (2)

Elementary lesions are mitral valve supra-ring, mitral stenosis, aortic stenosis and aortic coarctation. Our patient has an obstruction at 3 levels: mitral stenosis, bicuspid aortic valve with aortic stenosis and isthmic coarctation. Mitral valve obstruction with isolated left ventricular outflow tract obstruction represents a more severe form of Shone's complex. (3)

The primum movens, which happen in the early embryogenesis period, is the mitral obstruction, which triggers the poor development of the left ventricular cavity leading to obstructions in the way of left heart ejection and possibly a coarctation of the aorta. By the development of prenatal diagnosis techniques, it is possible to make the diagnosis by the fetal ultrasound but it is usually misleading. In our case, the diagnosis was missed on pre-natal ultrasound. The complex can be diagnosed early in the neonatal period but it is more often made later according to the form and the degree of obstruction. There is a description of a form fruste of Shone's complex in 65 year-old man. (4) In our case, the diagnosis was made early at the age of 57 days. Clinical manifestations are related to the age and the degree of obstruction and include refusal of feeds, cough, dyspnea, edema of extremities, cyanosis, hypertension and heart failure signs. Our patient presented with respiratory distress, cyanosis and heart failure signs. The diagnosis is confirmed by transthoracic echocardiography. CT angiography is useful to determine precise numbers, positions and morphologies of obstruction levels. Shone's complex need urgent surgical correction and interventions several times. Mitral valve repair is indicated afterwards because it became clinically significant in childhood and early reparation is associated with higher incidence of restenosis. The intervention should be considered before the occurrence of pulmonary hypertension. (5) Patients have been operated at varying time frames (2 days-3 years of age). (6) Our patient was operated on day 60 of life.

The prognosis is related to the severity of mitral involvement, the degree of pulmonary hypertension and the surgical treatment option. (5) The mortality increases to 24% in the second intervention. (7)

To conclude, the finding of aortic coarctation should prompt for search of other obstructive cardiac defects on the left heart. The prognosis is related to the degree of mitral stenosis and surgical treatment options.

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