Le complexe de Shone atypique à propos d'un cas
Atypical Shone's complex: Pediatric case report

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Résumé :

Le complexe de Shone est une cardiopathie congénitale extrêmement rare, une cinquantaine de cas ont été rapportés dans la littérature. Elle est caractérisée par une grande variabilité dans l'expression clinique et pose des problèmes de diagnostic positif, de prise en charge et de pronostic. Le complexe de Shone correspond à une association de malformations obstructives du cœur gauche à plusieurs niveaux. Le complexe de Shone complet ou typique dans sa description première, correspond à 4 niveaux d'obstruction et associe une membrane supra-valvulaire mitrale, une sténose de la valve mitrale due à une valve mitrale en parachute, un obstacle sub-aortique et une coarctation de l'aorte. Depuis, plusieurs descriptions de syndrome de Shone à trois ou à deux niveaux d'obstruction ont été rapportés réalisant ainsi un complexe de Shone incomplet ou atypique.
Nous rapportons l'observation d'un nourrisson de 57 jours présentant un complexe de Shone atypique. Il a été opéré à l'âge de 60 jours avec une évolution favorable.

Abstract :

Shone's complex is an extremely rare and severe congenital heart disease, until now about fifty cases are reported in the literature. It is characterized by great clinical variability and difficulties in diagnosis and management. Shone's complex consists on the association of obstructive defects on 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 and associated 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. The prognosis of children is still generally poor. We report the case of a 57-day-old boy transferred to pediatric department for persistant 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 favourable outcome.

Shone's complex is a rare condition, first described by Shone in 1963 [1]. It consists in 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.

Mots clés : enfant, complexe de shone ; membrane supravalvulaire mitrale ; sténose mitrale ; coarctation de l'aorte.
Keys word: Child; Shone's complex; Supramitral ring; Mitral stenosis; Aortic coarctation

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CASE PRESENTATION

A 57-day-old male infant was referred to our department for management of dyspnea. He is born to a 32-year-old mother primigravida with no significant history. He was derived from a first degree consanguineous marriage. The pregnancy was uneventful and prenatal ultrasounds were normal. He was born by means of normal vaginal delivery. 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 dyspnea with feeding difficulties and was admitted to our department.

On physical examination, the weight was 3770g. He was hypothermic. He has respiratory distress with tachypnea and retraction signs. The extremities were cold. He was cyanotic with oxygen saturation at 85% in the air. The cardiac auscultation revealed a galloping sound. The liver was enlarged at 6 cm. Femoral pulses were not found contrasting with the presence of axillary pulses suggesting aortic coarctation.

On the chest X-ray there was a normal cardiac silhouette and a bronchoalveolar syndrome. The electrocardiogram showed a left ventricular hypertrophy.

The 2-D transthoracic echocardiogram 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 8mmHg. Aortic valve was bicuspid with aortic stenosis. There was a left ventricular hypertrophy. The maximum gradient between the left ventricle and the aorta is estimated at 19 mmHg. This gradient is underestimated because of the coarctation of the aorta. The pulmonary arterial pressures were normal.

Computed tomography angiography was performed and showed the aortic coarctation and the left ventricle hypertrophy (figure 1).

Surgical coarctation repair was performed in the 60th day of life. The correction of mitral stenosis will be planned in a second time. The postoperative course was uneventful.

Figure 1 : Computed tomography angiography showed the aortic coarctation.

DISCUSSION

Shone's complex is an extremely rare congenital heart disease described by Shone in 1963 [1]. Zucker et al. found 4 cases when they reviewed 12520 prenatal ultrasounds [2]. The primum movens, which happen in the early embryogenesis period, is the mitral obstruction, 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, prenatal ultrasound was normal.

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 [3]. 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 included refude feeding, cough, dyspnea, edema of extremities, cyanosis, hypertension, heart failure signs. Our patient presented respiratory distress, cyanosis and heart failure signs.

The diagnosis is confirmed by transthoracic echocardiography. CT angiography is useful and precise numbers, positions and morphologies of obstruction levels.

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% [4]. 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.

Shone's complex need urgent surgical correction that may require intervention on several times. Mitral valve repair is indicated on the second time and should be considered before the occurrence of pulmonary hypertension [5]. The mean age at the first intervention was 9months (2 days-3 years) [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]. Thus, the mortality increases to 24% in the second intervention [7]. For the most severe cases, the prognosis continues to be poor [8]. Mitral valve obstruction with isolated left ventricular outflow tract obstruction represents a more severe form of Shone's complex [8].

LEARNING POINTS/TAKE HOME MESSAGES

- The rarity and variety of heart obstructive lesions observed in the atypical Shone complex explain the difficulty of the diagnosis.
- Diagnosis of Shone's syndrome by fetal echocardiography at mid-gestation may be misleading, taking into consideration the fact that the fetal heart is still actively growing and developing and the diagnosis is usually made after birth by echocardiography
- 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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