Case report

Aortic atresia with aortico-left ventricular tunnel mimicking severe aortic incompetence in utero

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Abstract

A fetus was diagnosed with regurgitation from the aorta into the left ventricle. A large aortico-left ventricular tunnel with aortic atresia was identified postnatally by echocardiography in a neonate suffering from severe heart failure. We successfully reconstructed the valve and closed the tunnel with a patch on his first day of life. At three-month follow-up, the baby is asymptomatic with mild aortic regurgitation. The combination of congenital aortico-left ventricular tunnel and aortic atresia can be misinterpreted in prenatal echocardiography as aortic regurgitation. This pathology may require urgent operative interventions.

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1. Introduction

Aortico-left ventricular tunnel (ALVT) is rare congenital cardiac defect originally reported by Edwards in 1961 [1]. In this pathology, the tunnel bypasses the aortic valve via a paravalvular connection from the left ventricle to the aorta. From 130 reported cases, surgical repair was possible with acceptable mortality and promising long-term outcome (Orphanet encyclopedia, 2005; http://www.orpha.net/data/patho/GB/uk-AortoVentricularTunnel.pdf). Aortic lesions are frequently observed associated defects [2]. If the aortic valve is obstructed or dysplastic, it is difficult to differentiate prenatally such lesions from aortic valve incompetence. We report on a patient whose postnatal hemodynamics with severe cardiac failure made emergency repair necessary.

2. Case report

A fetus was diagnosed with regurgitation from the aorta to the left ventricle in gestation week 28. The neonate developed postnatal symptoms of severe heart failure requiring mechanical ventilation. We diagnosed an aortico-left ventricular tunnel with aortic atresia by echocardiography (Fig. 1). Surgical repair was performed on the first day of life on an emergency basis. The inspection of the heart revealed dilated great arteries and an aneurysmatic aortico-left ventricular tunnel at the anterior LV wall (Fig. 2). After initiation of CPB and under cardiac arrest, we opened the tunnel longitudinally and performed aortotomy to directly inspect these findings. The tunnel’s aortic end arose from the right coronary sinus, with the right coronary orifice at its entrance. The aortic valve was bileaflet and the leaflets were thickened, myxomatous, and fused. The orifice was obstructive, allowing no intubation with a thin probe.

We closed the tunnel with a PTFE patch at the aortic entrance distally to the right coronary orifice. The valve repair was performed by dissecting the two commissures from the aortic wall up to the aortic annulus. Thereafter, the two commissures were divided to create an orifice of 8—10 mm in diameter. The tunnel and aortotomy were closed with running sutures. Intraoperative echocardiography showed mild aortic regurgitation. By intraoperative angiogram, we verified definite tunnel closure. Ventricular function was maintained with mild inotropic support. The postoperative course was uneventful. On three-month follow-up, the child was asymptomatic and receiving no oral medication. Echocardiography shows mild aortic regurgitation with normal LV function and dimensions.

3. Discussion

Congenital aortico-left ventricular tunnel is a rare cardiac malformation frequently associated with aortic lesions [3,4]. If the aortic valve is obstructed, this pathology’s prenatal diagnosis can be difficult to differentiate from aortic valve regurgitation. Furthermore, the postnatal hemodynamic changes with massive regurgitation volumes can necessitate
urgent treatment. Therefore, prompt, postnatal echocardiographic diagnosis in specialized centers is necessary in such cases.

Surgical repair is believed to be the proper treatment for ALVT with good late functional results anticipated [2]. Neonates with large tunnels and associated aortic valve obstruction might require urgent operative intervention to prevent rapid worsening of heart failure, as in our case. In this scenario, aortic valve reconstruction is preferable to avoid or delay valve replacement. However, a homograft implantation is an alternative treatment option if the graft is available in the urgent setting. Finally, the Ross procedure can be performed as a last resort but that is associated with a higher risk in the neonate [5].

4. Conclusions

The rare finding of ALVT and aortic valve atresia mimics aortic valve regurgitation in utero. These associated pathologies might require urgent surgical repair. In the present case, the tunnel closure and aortic valve reconstruction were carried out successfully and we anticipate good long-term results.

Fig. 1. Prenatal echocardiography (Section 2 according to Chaoui): visualization of left ventricular (LV) hypertrophy with systolic antegrade blood flow in the aorta (Ao) in the left picture and diastolic severe regurgitation in the right. In retrospect, the atretic valve was hidden behind the hugely dilated aortico-left ventricular tunnel.

Fig. 2. Intraoperative findings: note the aneurysm near the right ventricular outflow tract representing the aortico-left ventricular tunnel (left picture). The tunnel is opened longitudinally (right picture). The atretic aortic valve can be seen directly next to the PTFE-patch that was used to close the tunnel.
References


