Case Report

MULTIDISCIPLINARY MANAGEMENT OF A RARE CASE OF MIXED TOTAL ANOMALOUS PULMONARY VENOUS CONNECTION

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Running title. Rare case of mixed TAPVC

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Abstract

A rare case of mixed total anomalous pulmonary venous connection (TAPVC) associated to right extra-lobar bronchopulmonary sequestration (BPS) was diagnosed at birth in a full-term newborn. At one month of age, the patient underwent embolization of the BPS, complicated by coil entrapment in the right common iliac artery requiring urgent laparotomy. Few days later, the congenital cardiac repair was accomplished uneventfully.

At 12-months follow-up, the patient did not have pulmonary hypertension, but presented a moderate stenosis of the right femoral artery, which was effectively treated with anticoagulation therapy.

Keywords

Total Anomalous Pulmonary Venous Connection, Bronchopulmonary Sequestration, Transcatheter Embolization, Amplatzer Plug

Introduction

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This paper reports the case of an infant affected by mixed total anomalous pulmonary venous connection (TAPVC), an extremely rare congenital heart disease (CHD) (1), and bronchopulmonary sequestration (BPS), representing the second most frequent lung malformations in children (2). We described our multidisciplinary therapeutic strategy of a complication occurred during the embolization of the anomalous BPS vessel and discussed possible different surgical approaches to correct both the TAPVC and BPS.

Case presentation

A full-term male newborn, with hypoxemia at birth, was diagnosed with mixed TAPVC, in which the left pulmonary veins (PVs) were draining into the coronary sinus and the right PVs were connected to the inferior vena cava (Scimitar-syndrome).

The cardiac catheterization also described a right extralobar BPS, supplied by a vessel from the celiac artery (*Figure 1*).

A multi-stage elective treatment was planned which included the percutaneous embolization of the anomalous vessel at the age of 1 month, to prevent pulmonary systemic overflow and pulmonary hypertension, followed by the repair of the CHD.

Through a 4Fr Benson® catheter, a 5-mm-Amplatzer® vascular plug was placed in the anomalous vessel. A second 5x5-mm-Cook® coil was inserted but remained entrapped in the celiac artery. Attempting to remove it, the coil remained blocked in the right common iliac artery (*Figure 2*). An urgent laparotomy was performed through a Rutherford-Morrison incision. The iliac artery was incised, the coil was removed, and 2Fr Fogarty catheter angioplasty allowed to remove a femoral artery thrombus.

On POD 1, a moderate stenosis of the right femoral artery was detected, due to possible clotting. Systemic urokinase was administered for 48 hours, followed by continuous infusion of heparin until the next surgery.

Seven days after, the patient underwent partial cardiac repair, consisting in baffling the left anomalous pulmonary venous flow into the left atrium, unroofing the coronary sinus and closing the coronary sinus ostium with a patch. The child was discharged on POD 26 on warfarin and metoprolol due to postoperative supraventricular tachycardia.

At 12-months follow-up, the patient is growing well, with no recurrence of arrhythmias. Femoral arterial flow was improved by collateral circles. Cardiac evaluation showed good ventricular function, with no residual pulmonary hypertension and good left pulmonary venous flow. Metoprolol and warfarin were discontinued after six months and the patient was scheduled for the final cardiac repair at school age.

Comment

Repair of complex TAPVD is challenging and presents a high risk of postoperative pulmonary hypertension and possible occurrence of stenosis (3), which raises the overall mortality up to 40% (1). The extralobar BPS is normally located at the base of the left chest, and children may be asymptomatic or have important respiratory distress and severe congestive heart failure secondary to the volume-loading from a large systemic artery for the sequestered segment (2). The thoracoscopic resection of the lesion is the standard treatment.

These anomalies require the involvement of pediatric cardiologists pediatric surgeons and cardiothoracic surgeons, in a tertiary-care center, who share different skills for defining the diagnostic/therapeutic approaches (4).

The BPS embolization using the Amplatzer device was indicated to prevent postoperative pulmonary overflow, as already described (5). However, large case series of BPS embolization showed a high rate of failure, due to incomplete embolization, and persistence of the anomalous lung tissue (2).

A further issue concerns the observed complication. This could be resolved through an extraperitoneal approach, normally used for kidney transplantations.

Conclusion

The multidisciplinary approach allowed a successful treatment of these complex anomalies, despite the potential complications of BPS embolization.

The multidisciplinary team should be aware that BPS may persist after embolization, and possible alternative methods of treatment should be discussed

Author contribution

Concept/design: FG, MP, PD

Drafting: FG, MP, PD

Critical revision: BC, MP, PD

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Legends to figures

Figure 1. First cardiac catheterization; right anomalous pulmonary venous connection (a); anomalous arterial vessel from the celiac artery (b).

Figure 2. Second cardiac catheterization; positioning of the Amplatzer plug (a, b); entrapment of the coil (c).