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CASE REPORT

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Left extralobar pulmonary sequestration and a right aorto-to-pulmonary vein fistula in a newborn: a 3-mm thoracoscopic monolateral approach

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Abstract

An extralobar pulmonary sequestration (EPS) associated with a contralateral aorto-to-pulmonary vein fistula is rare. We report the case of a female newborn with left EPS fed by an artery originating from the distal thoracic aorta and, symmetrically on the controlateral side, an artery shunting in the inferior right pulmonary vein. Echocardiography showed dilatation of the left atrium. On the 34th day since birth (weight 4500 g), the patient was operated on thoracoscopically. The EPS was closed with a 3-mm sealing system, divided and removed. A window in the mediastinal pleura was created, and the origin of the fistula was identified and sealed. The postoperative course was uneventful. The patient was discharged on Day 4 with no echocardiographic signs of persistence of the fistula and of the congestive heart failure. This is the first case report of a thoracic large systemic circulation-to-pulmonary vein fistula causing heart failure associated with EPS. The thoracoscopic monolateral approach and the availability of 3-mm instruments guaranteed a maximum level of minimal invasiveness.

Keywords: Pulmonary sequestration • Aorto-to-pulmonary vein fistula • Thoracoscopy • Newborn

INTRODUCTION

Cases of an isolated aorto-to-pulmonary vein fistula have been previously described both in children and adults. However, an extralobar pulmonary sequestration (EPS) associated with a contralateral aorto-to-pulmonary vein fistula is rare [1].

CASE REPORT

A female newborn with prenatal ultrasonographic and magnetic resonance imaging (MRI) suspicion of left pulmonary sequestration (34 × 45 × 40 mm) was delivered at our centre (gestational age 39 weeks, birth weight 3.410 g). Gestation was uneventful, and the suspicion was raised during the morphological ultrasound in the second trimester. After birth, MRI without contrast and sedation confirmed a left pulmonary sequestration fed by a 3-mm arterial vessel originating from the anterior wall of the distal thoracic aorta and, symmetrically on the right side of the aorta, a 2.8-mm artery shunting in the inferior right pulmonary vein. It also showed a vein draining the sequestration into the right inferior pulmonary vein. Echocardiography showed dilatation of the right pulmonary vein and left atrium. An angiocomputed tomography (CT) scan confirmed the vascular malformations (Fig. 1). Left heart dilation and Pro-BNP values of 23.215 pg/ml (normal value 0–450 pg/ml) despite the diuretic therapy (furosemide 1 mg/kg/day) prompted surgery. On the 34th day after birth (weight 4500 g), the patient was operated on, and tracheal intubation, a right lateral position, three-mm trocars (1 in the 4th intercostal space below the tip of the scapula and 2 in the midaxillary line in the 2nd and 5th intercostal spaces) and 4-mmHg pneumothorax were chosen. An EPS was observed, and its vessels were closed with a 3-mm sealing system (Just-Right Surgical®), divided and removed. A window in the mediastinal pleura was created, and the origin of the right aorto-to-pulmonary vein fistula was found to be symmetrical to the origin of the previously divided artery. The fistula was sealed with the JustRight Surgical® system close to its origin (Fig. 2). Pathology confirmed the diagnosis. The postoperative course was uneventful. The patient was discharged on Day 4 with no echocardiographic signs of a persisting fistula. Diuretic therapy was stopped 2 weeks after surgery. Pro-BNP values returned to normal.

DISCUSSION

An abdominal aorta-to-pulmonary vein fistula is a rare congenital anomaly previously described in infancy and adulthood [1]. To the best of our knowledge, this is the first case report of a thoracic systemic circulation-to-pulmonary vein fistula associated with EPS. The differential diagnosis includes scimitar syndrome,

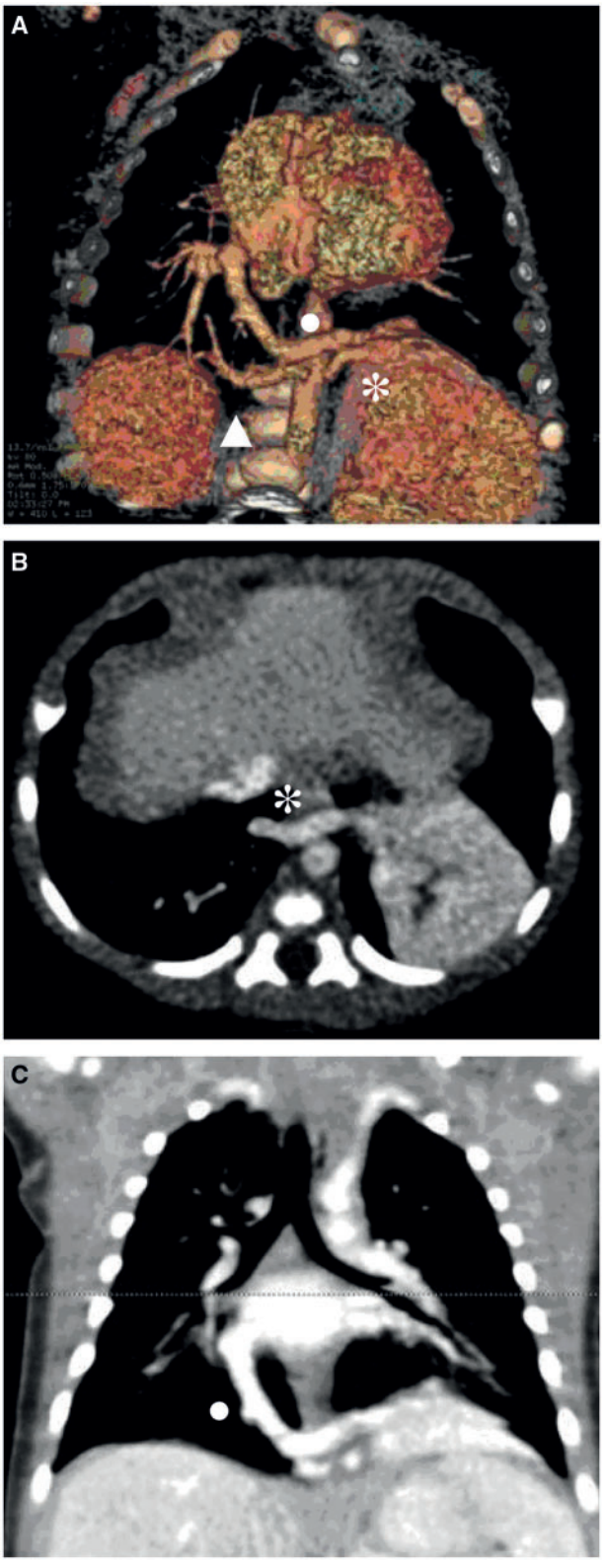

Figure 1: (**A**) Magnetic resonance imaging: *, arterial; •, venous vessels of the sequestration and ▲, aorto-to-pulmonary vein fistula. (**B**) An angiocomputed tomography scan: *, the aortic origin of the 2 arterial branches. (**C**) An angiocomputed tomography scan: •, venous drainage of the sequestration.

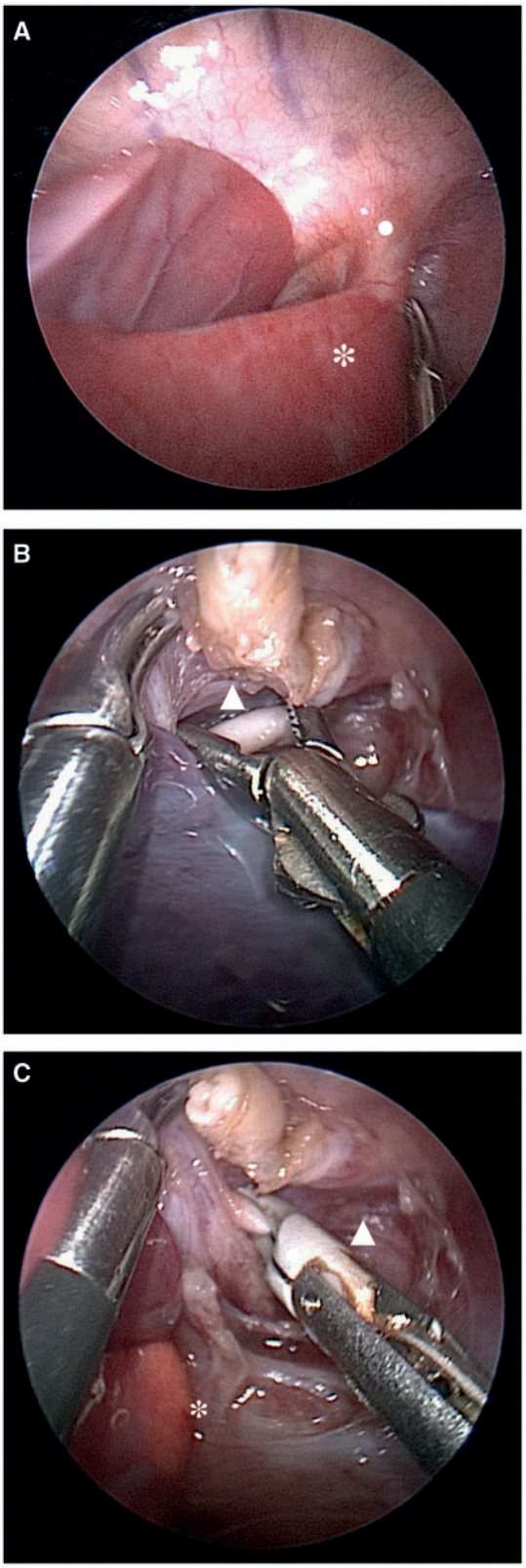

Figure 2: (**A**) *, Extralobar pulmonary sequestration and •, artery feeding the sequestration. (**B**) ▲, Aorto-to-pulmonary vein fistula. (**C**) ▲, Sealing the aorto-to-pulmonary vein fistula.

with a communication between the pulmonary vein and vena cava, which is sometimes associated with pulmonary sequestration [2, 3]. The multimodality imaging allowed an accurate anatomical definition, and is thus extremely useful to define the right surgical approach. Lung sequestration is defined as a non-functioning mass of normal lung tissue lacking a normal communication with the pulmonary arterial system and the tracheobronchial tree and receiving the blood supply from 1 or more anomalous systemic arteries. In the presence of a distinct pleural covering and a complete anatomical separation of the mass from adjacent normal lung tissue, the sequestration is defined as extralobar and is usually managed by resection. An aorta-to-pulmonary vein fistula without sequestration can be managed by surgical ligation or percutaneous closure of the fistula [4]. In recent years, thoracoscopy has emerged as an established technique for the management of congenital pulmonary malformations in infancy [5] and has also been proven to be a safe and effective procedure in the treatment of this rare and complex congenital malformation. The monolateral approach and the availability of 3-mm instruments guaranteed a maximum level of minimal invasiveness.

Conflict of interest: none declared.

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