Scimitar syndrome and left pulmonary vein stenosis: A serious and rare association

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ABSTRACT

Scimitar syndrome is a rare constellation of cardiovascular anomalies consisting of partial right anomalous pulmonary venous drainage to the inferior vena cava (IVC), hypoplasia of the right lung, cardiac dextroposition, and in many cases, one or more aortopulmonary collaterals from the descending aorta to the hypoplastic lung. The “infantile form” commonly presents with significant heart failure and pulmonary hypertension. Stenosis of the left pulmonary veins associated with scimitar syndrome is a rare but well-described combination, usually associated with a poor prognosis. We describe two patients with this association in whom the left pulmonary vein stenosis manifested months after the initial diagnosis as a progressive lesion that complicated the course of the disease due to severe pulmonary hypertension. Both patients were successfully treated with a combination of surgical, percutaneous, and hybrid treatment.

Keywords: Pulmonary hypertension, pulmonary vein stenosis, scimitar syndrome

INTRODUCTION

Scimitar syndrome is a rare cardiovascular anomaly consisting of partial right anomalous pulmonary venous drainage to the inferior vena cava (IVC), hypoplasia of the right lung, cardiac dextroposition, and in many cases, one or more aortopulmonary collaterals from the descending aorta to the hypoplastic lung. The presence of an atrial septal defect and signs of suprasystemic pulmonary hypertension. A cardiac catheterization was performed to assess pulmonary pressures which were 90% systemic with an elevated pulmonary-to-systemic flow ratio (Qp/Qs of 2). No aortopulmonary collaterals were found but a partial anomalous pulmonary venous drainage was suspected but not confirmed due to hemodynamic instability. A computed tomography (CT) scan revealed an anomalous drainage of the right lower pulmonary vein into the suprahepatic segment of the IVC, which was missed on the echocardiogram [Figure 1]. Due to the patient’s low weight (3.400 kg), surgery was not considered at this moment and the treatment performed on admission showed cardiac dextroposition with the presence of an atrial septal defect and signs of suprasystemic pulmonary hypertension. A cardiac catheterization was performed to assess pulmonary pressures which were 90% systemic with an elevated pulmonary-to-systemic flow ratio (Qp/Qs of 2). No aortopulmonary collaterals were found but a partial anomalous pulmonary venous drainage was suspected but not confirmed due to hemodynamic instability. A computed tomography (CT) scan revealed an anomalous drainage of the right lower pulmonary vein into the suprahepatic segment of the IVC, which was missed on the echocardiogram [Figure 1]. Due to the patient’s low weight (3.400 kg), surgery was not considered at this moment and the treatment performed on admission showed cardiac dextroposition with the presence of an atrial septal defect and signs of suprasystemic pulmonary hypertension. A cardiac catheterization was performed to assess pulmonary pressures which were 90% systemic with an elevated pulmonary-to-systemic flow ratio (Qp/Qs of 2). No aortopulmonary collaterals were found but a partial anomalous pulmonary venous drainage was suspected but not confirmed due to hemodynamic instability. A computed tomography (CT) scan revealed an anomalous drainage of the right lower pulmonary vein into the suprahepatic segment of the IVC, which was missed on the echocardiogram [Figure 1]. Due to the patient’s low weight (3.400 kg), surgery was not considered at this moment and the treatment performed on admission showed cardiac dextroposition with the presence of an atrial septal defect and signs of suprasystemic pulmonary hypertension. A cardiac catheterization was performed to assess pulmonary pressures which were 90% systemic with an elevated pulmonary-to-systemic flow ratio (Qp/Qs of 2). No aortopulmonary collaterals were found but a partial anomalous pulmonary venous drainage was suspected but not confirmed due to hemodynamic instability. A computed tomography (CT) scan revealed an anomalous drainage of the right lower pulmonary vein into the suprahepatic segment of the IVC, which was missed on the echocardiogram [Figure 1]. Due to the patient’s low weight (3.400 kg), surgery was not considered at this moment and the treatment performed on admission showed cardiac dextroposition with the presence of an atrial septal defect and signs of suprasystemic pulmonary hypertension. A cardiac catheterization was performed to assess pulmonary pressures which were 90% systemic with an elevated pulmonary-to-systemic flow ratio (Qp/Qs of 2). No aortopulmonary collaterals were found but a partial anomalous pulmonary venous drainage was suspected but not confirmed due to hemodynamic instability. A computed tomography (CT) scan revealed an anomalous drainage of the right lower pulmonary vein into the suprahepatic segment of the IVC, which was missed on the echocardiogram [Figure 1]. Due to the patient’s low weight (3.400 kg), surgery was not considered at this moment and the treatment

CASE REPORTS

Case 1

A 2-month-old infant was transferred to our facility due to severe pulmonary hypertension. An echocardiogram performed on admission showed cardiac dextroposition with the presence of an atrial septal defect and signs of suprasystemic pulmonary hypertension. A cardiac catheterization was performed to assess pulmonary pressures which were 90% systemic with an elevated pulmonary-to-systemic flow ratio (Qp/Qs of 2). No aortopulmonary collaterals were found but a partial anomalous pulmonary venous drainage was suspected but not confirmed due to hemodynamic instability. A computed tomography (CT) scan revealed an anomalous drainage of the right lower pulmonary vein into the suprahepatic segment of the IVC, which was missed on the echocardiogram [Figure 1]. Due to the patient’s low weight (3.400 kg), surgery was not considered at this moment and the treatment performed on admission showed cardiac dextroposition with the presence of an atrial septal defect and signs of suprasystemic pulmonary hypertension. A cardiac catheterization was performed to assess pulmonary pressures which were 90% systemic with an elevated pulmonary-to-systemic flow ratio (Qp/Qs of 2). No aortopulmonary collaterals were found but a partial anomalous pulmonary venous drainage was suspected but not confirmed due to hemodynamic instability. A computed tomography (CT) scan revealed an anomalous drainage of the right lower pulmonary vein into the suprahepatic segment of the IVC, which was missed on the echocardiogram [Figure 1]. Due to the patient’s low weight (3.400 kg), surgery was not considered at this moment and the treatment

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How to cite this article: Mendoza A, Herrera D, Caro AT, García E, Montañés E, Granados MA. Scimitar syndrome and left pulmonary vein stenosis: A serious and rare association. Ann Pediatr Card 2022:15:80-3.
consisting of diuretics and angiotensin-converting enzyme inhibitors was started. At the age of 6 months, the patient deteriorated in the setting of a respiratory infection. Clinical presentation was consistent with pulmonary hypertensive crisis requiring intubation. On echocardiogram, left pulmonary venous confluence stenosis was suspected. Further, cardiac catheterization showed a filiform stenosis at the IVC to right atrium junction, above the drainage of the lower right pulmonary vein [Figure 2a], and a stenosis of 2.1 mm in the left pulmonary venous confluence [Figure 2b], which was treated with a Maverick XL® 6 mm × 20 mm balloon dilation with initial satisfactory result. Attempted surgical repair of the anomalous right pulmonary venous drainage was technically challenging and hence not done. Instead, a patch enlargement of the IVC and partial closure of the atrial septal defect was performed. In the postoperative period, the patient continued to show signs of pulmonary hypertension and progressive stenosis of the left pulmonary veins. A third catheterization was performed at the age of 8 months, with a view to stent the left pulmonary venous confluence. Pulmonary pressure was 90% systemic. This procedure was complicated with a migration of the stent into the left atrium; therefore, the patient was transferred to the operating room for stent removal. In the same intervention, he underwent a hybrid procedure, with a stent implantation in the left pulmonary venous confluence (8 mm × 12 mm Formula® 418 stent), another in the IVC (7 mm × 12 mm Formula® 414 stent), and a third one in the right upper pulmonary vein (5 mm × 12 mm Formula® 418 stent), which also had a stenotic appearance. The patient was discharged home 2 months later. At the age of 14 months, he presented again with hemodynamic decompensation, pulmonary hypertension, and signs of recurrence of stenosis of the stented left pulmonary veins. At this time, another hybrid procedure was performed, showing a stenosis of the collector distal to the previously placed stent [Figure 3a], which resolved with the placement of a new stent [Figure 3b]. Subsequently, the clinical course was satisfactory. At his last clinical assessment at the age of 7 years, the patient had a good functional class and noninvasive imaging (echocardiogram and CT scan) revealed obliteration of the lower right pulmonary vein into the IVC connection with collateral circulation into the vascular territory of the right upper pulmonary vein and mild stenosis of the stents placed in the left pulmonary veins [Figure 4]. Mild pulmonary hypertension was seen on echocardiogram with a pulmonary systolic pressure (PSP) estimated by tricuspid regurgitation (TR) of 25 mmHg.

Case 2
A newborn patient with a prenatal diagnosis of scimitar syndrome with right lung hypoplasia, right pulmonary artery hypoplasia, and anomalous pulmonary venous drainage of this lung to the IVC was admitted to our neonatal unit. An echocardiogram also showed an ostium secundum atrial septal defect. A CT scan revealed a horseshoe lung [Figure 5] and two aortopulmonary collateral arteries arising from the descending abdominal aorta to the base of the right lung. At the age of 3 weeks,
a cardiac catheterization showed severe pulmonary hypertension with a pulmonary artery pressure at 80% of the systemic pressure and a Qp/Qs of 2.8. The two aortopulmonary collaterals were embolized during the procedure with a decrease in the pulmonary to systemic pressure down to 46% [Figure 6]. The patient improved clinically and was discharged home. However, at the age of 4 months, an echocardiogram showed an abnormal gradient in the drainage of the left pulmonary veins. A diagnostic cardiac catheterization demonstrated left pulmonary vein stenosis with a 20 mmHg pressure gradient between the left pulmonary venous confluence and the left atrium. Pulmonary artery pressure was 60% of the systemic one. With these findings, the patient underwent surgical intervention with direct anastomosis of the right pulmonary vein into the left atrium and repaired the left pulmonary vein stenosis with sutureless technique. Three months postoperatively, the patient required surgical reintervention due to stenosis of the left pulmonary venous confluence. Again, a sutureless technique was used. At the age of 4 years, the patient remained in a good functional class, with mild residual stenosis of the left pulmonary veins and PSP estimated by TR of 35 mmHg.

**DISCUSSION**

Scimitar syndrome is a rare constellation of cardiovascular anomalies. In infants, the disease typically presents with a multifactorial pulmonary hypertension due to (i) left-to-right shunt secondary to aortopulmonary collateral arteries, anomalous pulmonary venous drainage, and septal defects, (ii) right pulmonary hypoplasia. The association between stenosis of the left pulmonary veins and scimitar syndrome is an additional complicating factor for pulmonary hypertension in this context and is associated with a poor prognosis. Nine patients with this association have been described in the literature, eight with a fatal outcome and the remaining requiring a lung transplant.

This association suggests a generalized disorder of the pulmonary veins. Early in gestation, when the connection of the outpouching of the left atrium with the pulmonary venous plexus takes place, this would manifest as anomalous pulmonary venous drainage, and after birth with a progressive myofibroblastic proliferation which can explain why the left pulmonary vein stenosis was not present at birth but occurred months after the initial diagnosis.

Treatment of these patients is complex, as it involves correction of the anomalous pulmonary venous connection, the associated heart defects, in addition to the relief of the left pulmonary vein stenosis during infancy. Furthermore, recurrence of the left pulmonary vein stenosis is a common postoperative complication, which can require combined surgical, percutaneous, and hybrid treatment.
Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the forms, the patients have given their consent for their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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