A PARTICULAR INFANTILE SCIMITAR SYNDROME VARIANT WITH ANOMALOUS SYSTEMIC ARTERIAL SUPPLY-INFERIOR VENA CAVA FISTULA

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Short running title: A RARE VASCULAR ANOMALY

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Abstract

Scimitar syndrome is a rare anomaly, most commonly including partial pulmonary venous drainage into the inferior vena cava, hypoplasia of the right lung, and systemic collaterals from the aorta. We report a pediatric case in whom this rare syndrome was associated with intracardiac lesions and a large aberrant systemic arterial supply to the right lung that originated from the abdominal aorta and fistulated in the inferior vena cava.

Keywords: Scimitar Syndrome; pediatric; aorta-inferior vena cava fistula

Introduction

Simitar syndrome is a rare cardiovascular and bronchopulmonar congenital abnormality, the incidence ranges from 1:5/100.000 live births (1,2). The components of Scimitar syndrome include: partial pulmonary venous drainage into the inferior vena cava (IVC), variable degrees of hypoplasia of the right lung with rotation of the heart into the right hemitorax, hypoplasia of the right pulmonary artery (PA), and an aberrant systemic arterial supply from the aorta to the right lung (3). The syndrome is variable in its expression, variants of this congenital condition had been widely described (4,5,6). Additional intracardiac lesions are well recognised, with a prevalence of 40% (2).

Case report

We report the original case of a 6-weeks-old female infant who was transfered in our tertiar center, presenting acute dyspnea and requiring pediatric intensive care unit admission and intubation. She was born at 36 weeks of gestation with a birth weight of 3000g. The physical examination showed respiratory distress, signs of congestive heart failure, and the lower limbs presented intermittent coldness. Cardiac auscultation revealed tachycardia with systolic murmur. A chest radiography revealed opacification of the right hemithorax, cardiac enlargement with moderate displacement of the cardiac structures from left-to-right, pulmonary plethora; a scimitar vein was not recognized. Echocardiography showed mesocardia, dilatation of the IVC, dilatation of both ventricles with normal function, interatrial communication (5mm) ostium secundum type with left-to-right shunt, ventricular septum defect (4mm) muscular type, hypoplasia of the right PA (z score: -2.4). Only the left pulmonary veins were recognized by echocardiography on the left side, a right pulmonary vein drained into the right atrium (RA) and a small pulmonary vein drained into the IVC (*Figure 1A,B*). The subcostal view revealed a large artery vessel arrising from the abdominal aorta near the celiac axis, with ascending and tortuos course (*Figure 1C,D*).

CT-scan revealed hypoplasia of the right lung, with right PA hypoplasia; no parenchyma appeared excluded from normal bronchial tree (*Figure 2A*). An anomalous venous return was observed: a right dilated pulmonary vein opening into the RA drained the pulmonary venous circulation corresponding to the right upper and middle lobe, and the upper and anterobasal segments of the right lower lobe (*Figure 2B*); another pulmonary vein collecting laterobasal and posterobasal segments of the right lower lobe crosses the diaphragm and joins with another pulmonary vein that drains the mediobasal segment of the right lower lobe, than draining together in the IVC. There was a large aberrant artery arrising from abdominal aorta above the celiac trunk with ascending, transdiaphragmatic course, which separeted in two branches: a branch supplying the lower part of the right pulmonary circulation, and the other branch draining in the cranial portion of the IVC, representing an aorta-IVC fistula (*Figure 2C,D*). Below the origin of this artery the aorta was narrowed.

Cardiac catheterization confirmed the diagnostic of Scimitar variant, with the presence of systemic-to-pulmonary collateral vessel arrising from abdominal aorta below the diaphragm, which supplied an important area of the right lung and from which an aorta-venous fistula has developed, draining into the IVC (*Figure 3*). The hemodynamics revealed a mean PA pressure of 32mmHg, Qp:Qs=7.82, Rp:Rs=0.2, oxygen saturation of 55.8% in IVC at the level of the lower margin of the liver, and oxygen saturation of 94.4% in IVC at the level of RA.

In spite of vigorous treatment of congestive heart failure, the condition of the infant deteriorated. The patient underwent surgery on the 12^{th} day after admission to supress flow through the aberrant vessel. The anomalous vessel was identified and ligated immediately after its emergence, on cardiopulmonary by-pass. After this procedure there was an improvement in the infant`s condition. Postoperative echocardiography estimated a normal pressure in pulmonary circulation and demonstrated left-to-right shunt at three levels: at atrial and

ventricular level, and at the partial anomalous pulmonary venous return, with a Qp:Qs of 1.8:1. The anomalous venous return will be surgically treated later in evolution.

Publication of this case was approved by the institutional review board in the presence of a signed informed consent obtained from the mother.

Discussions

The syndrome was classified by Depuis et al. into two distinct forms: ”infantile” and ”adult” (3). The ”infantile” form affects children younger than one year and it typically has features of pulmonary arterial hypertension and congestive heart failure secondary to the: 1).significant left-to-right shunt from the anomalous pulmonary venous drainage, from an additional cardiac defect, and from the systemic arterial supply to the right lung, 2).reduction of the pulmonary vascular bed on the right side (1,2). The ”adult” form, without pulmonary hypertension, affects both children and adults, and is well tolerated and has a better prognosis (3).

The management of Scimitar syndrome is variable due to the variability in its clinical and hemodynamic expression. The operative approach should be individualized; a two-stage approach has been recommended (3). Embolisation through a catheter or surgical ligation of the aberrant arterial supply as the first stage showed clinical improvement of congestive heart failure by reduction of left-to-right shunt (1,2,7,8).

In our case, ligation of the anomalous systemic arterial collateral immediately after its origin reduced the left-to-right shunt by the elimination of the right pulmonary circulation supply and by the removal the aorta-IVC fistula.

The unique pattern of the presented case can be explained in the light of knowledge of the embryology. In early fetal life, there is a intercommunication between developing cardiac chambers and systemic, pulmonary, and venous systems. The persistence of communication of

the splahnic plexus with a cardinal or umbilicovitelline vein may lead to the appearance of some type of anomalous venous connection (9). If early connections of both arteries and veins persist, a congenital arteriovenous fistula may result. Le Rochais JP et al. reported a Scimitar syndrome associated with pulmonary arteriovenous fistulas (10). According to author`s knowledge, this is the first case published in the literature which associates this particular anatomical features, consisting in the association of the aorta-IVC fistula in a patient with Scimitar syndrome without pulmonary sequestration, association that makes this case unique.

Conclusions

Infantile Scimitar syndrome is a rare congenital anomaly that needs a high degree of suspicion and is difficult to diagnose and manage. Our case represents a unique variant of the Scimitar syndrome with normal bronchial tree, associated with intracardiac lesions and a large systemic arterial supply to the right lung, that originated from the abdominal aorta and also fistulated in the IVC.

Author contributions

All authors fulfill the criteria for authorship:

Carmen C Șuteu-data collection, data analysis/interpretation, drafting article

Cristina Blesneac-data collection, data analysis/interpretation

Marian Pop-data collection

Amalia Făgărășan-critical revision of article

Rodica Togănel-critical revision of article, approval of article

References

1. Depuis C, Charaf LA, Breviere GM, About P. “Infantile” form of Scimitar syndrome with pulmonary hypertension. Am J Cardiol 1993;71:1326-1330.

2. Rukban HAI, Ghaihab MAI, Tamini O, Al-Saleh S. Clinical spectrum of infantile scimitar syndrome: A tertiary center experience. Ann Pediatr Cardiol 2014;7(1):29-33.
3. Dupuis C, Charaf LA, Breviere GM, Abou P, Remy-Jardin M, Helmius G. The “adult” form of the Scimitar syndrome. Am J Cardiol 1992;70:502-507.
4. Argueta-Morales IR, Meador LC, Nykanen DG, DeCampli WM. Infantile form of scimitar syndrome with collateral pulmonary vein stenosis. Pediatr Cardiol 2010;31:550-552.
5. Babazadeh K, Zamani H, Ghaemi H, Omran FA. A case of variant scimitar syndrome. Caspian Journal of Pediatrics 2017;17(3):241-244.
6. Bo I, Daubeney PEF, Rigby ML. Uncommon variants of the Scimitar syndrome in two siblings. Annals of Pediatric Cardiology 2015;8:56-58.
7. Uthaman B, Abushaban L, Al-Qbandi M, Rathinasamy J. The impact of interruption of anomalous systemic arterial supply on scimitar syndrome presenting during infancy. Catheter Cardiovasc Interv 2008;71:671-678.
8. Brown JW, Ruzmetov M, Minnich DJ, Vijay P, Edwards CA, Uhlig PN, et al. Surgical management of Scimitar syndrome: An alternative approach. Journal of Thoracic and Cardiovascular Surgery 2003;125:238-245.
9. Ghandour A, Partovi S, Karappasamy K, Rajiah P. Congenital anomalies of the IVC-embriological perspectives and clinical relevance. Cardiovasc Diagn Ther 2016;6(6):482-492.
10. Le Rochais JP, Icard P, Davani S, Abouz D, Evrard C. Scimitar Syndrome with pulmonary arteriovenous fistulas. Ann Thorac Surg 1999;68:1416-8.