Supracardiac anomalous pulmonary venous connection with unilateral pulmonary venous atresia: Diagnosis and management

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ABSTRACT

We report a case of a 6-day-old neonate referred to us for surgical correction of total anomalous pulmonary venous connection. Meticulous evaluation contributed to accurate diagnosis of associated unilateral pulmonary venous atresia. This unique association provides insights into the importance of evaluation of all pulmonary veins using various imaging tools.

Keywords: Echocardiography, differential pulmonary venous congestion, multislice computed tomography

DOI: 10.4103/0974-2069.58318

INTRODUCTION

Anomalous pulmonary venous connection and drainage are known congenital cardiac anomalies that need early surgical correction. The presence of pulmonary venous stenosis or atresia in this setting may change management options. Thus, their coexistent presence should be specifically looked for during echocardiography.

This case emphasizes the importance of thorough evaluation of individual pulmonary veins before cardiac surgery. Multislice cardiac computed tomography, when indicated, may play a vital role in such an evaluation.

CASE REPORT

A 6-day-old, 2.8 kg full-term neonate with a history of fast breathing and feeding difficulty was referred to our hospital for surgical correction of supracardiac total anomalous pulmonary venous connection.

On examination, the baby had severe respiratory distress and central cyanosis with saturation of 72% by a pulse oximeter. Cardiovascular examination demonstrated a normal first heart sound, wide and fixed split second heart sound and a 2/6 ejection systolic murmur at the pulmonary area. Mechanical ventilation was started along with inotropes. Chest X-ray film showed haziness along with reticulogranular pattern involving the entire right lung. The left lung field was normal [Figure 1]. In retrospect, this pattern was suggestive of differential pulmonary venous congestion.

Transthoracic echocardiography showed left-sided anomalous supracardiac pulmonary venous connection. Left-sided pulmonary veins formed a common chamber behind the left atrium. From there, a right vertical vein ascended and joined the right innominate vein – superior vena cava junction, with a gradient of 11 mmHg at this junction. In addition, a large ostium secundum type of atrial septal defect (shunting right to left) was noted.

Figure 1: Chest X-ray film of the sick neonate on a mechanical ventilator – right lung shows reticulogranular pattern due to severe pulmonary edema. This differential pulmonary venous congestion is an important bedside clue to suspect unilateral pulmonary venous hypoplasia/atroresia.

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to left) and a small patent ductus arteriosus (shunting bidirectional) were observed. Right-sided pulmonary veins were not clearly seen.

Because the right veins were not clearly defined on transthoracic echocardiography, 64 multislice cardiac computed tomography was performed, which revealed atretic right-sided pulmonary veins with anomalous left-sided pulmonary venous connections and drainage, as described above [Figure 2]. Severe interstitial edema of the right lung was observed.

The case was discussed at our combined Pediatric Cardiology–Cardiac Surgical meeting. Surgical rerouting of unilateral pulmonary veins without addressing right lung issues may lead to persistence of severe pulmonary arterial hypertension (PAH). One option was to transplant the right lung and to reroute the pulmonary veins on the other side. Non-availability of an expert neonatal lung transplantation facility and “donor neonatal lung” along with very high procedural morbidity and mortality precluded such a repair in the neonatal period. Thus, this baby was deemed to be a poor candidate for cardiac surgery.

**DISCUSSION**

Although rare, total anomalous pulmonary venous connection is one of the routine congenital cardiac anomalies seen at tertiary care pediatric cardiac facilities. In most cases, all four pulmonary veins drain into a common chamber that has a variable drainage site. Pulmonary venous atresia is extremely rare with anomalous pulmonary venous connections but can affect management decisions. Etiology of pulmonary venous atresia is believed to be congenital and is related to a somewhat more common focal pulmonary venous stenosis.\(^{[1]}\) Improper incorporation of the common pulmonary vein into the left atrium is the embryological basis.

In a study by Makoto et al., nine of 48 patients of anomalous pulmonary venous connection had dysmorphic pulmonary veins. These included excessive tributaries, vertical vein atresia, hypoplastic confluence and focal pulmonary vein stenosis.\(^{[1]}\) None had pulmonary venous atresia involving the entire lung.

Older children with isolated unilateral pulmonary venous atresia present with hemoptysis, recurrent pulmonary infections and small lungs with reticulogranular pattern on a chest X-ray film.\(^{[2,5]}\) Venous drainage of the involved lung occurs into the azygos vein via the bronchial venous system.\(^{[3]}\) Treatment of this condition is lobectomy or pneumonectomy.\(^{[2,5]}\) Isolated pulmonary venous atresia is rare and its association with other congenital heart diseases is even rarer.\(^{[2]}\)

Evaluation of individual pulmonary veins is mandatory in a case with anomalous pulmonary venous connections. Takayuki et al. showed that the prospective detection rates of pulmonary venous abnormalities with echocardiography were 38%.\(^{[6]}\) Doubtful pulmonary venous anatomy on echocardiography can be further evaluated by cardiac computed tomography scan, which is an excellent diagnostic tool to define the pulmonary venous tree.\(^{[7,8]}\)

In a similar single reported case of infradiaphragmatic pulmonary venous connection and unilateral pulmonary venous atresia, the authors did not operate on their patient and mentioned unfavorable prognosis of such an entity.\(^{[9]}\)

In summary, early presentation, discrepancy between clinical condition and degree of pulmonary venous obstruction along with differential pulmonary venous congestion on chest X-ray film are important bedside clues to look for pulmonary venous hypoplasia/atresia, such as diagnosed in our case above. Unilateral small pulmonary artery (if present) is another clue for pulmonary venous hypoplasia/atresia, which is in contrast to dilated pulmonary arteries of classic total anomalous pulmonary venous connection.\(^{[10]}\)

Pre-surgical evaluation of individual pulmonary veins in congenital cardiac cases, especially those with anomalous pulmonary venous connections, prevents errors in management. Multislice cardiac computed tomography scan plays a vital role in accurate diagnosis of this rare entity.
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Source of Support: Nil, Conflict of Interest: None declared