

Case report

Scimitar syndrome: A rare cause of respiratory distress in infants – Case report

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Abstract

Scimitar syndrome is a rare combination of congenital cardiopulmonary anomalies that includes a partially anomalous right pulmonary vein (PARPV) from the right lung. Here we report an infant who presented with recurrent respiratory tract infections, respiratory distress and persistent pulmonary hypertension who was diagnosed to have scimitar syndrome. The diversity of the clinical presentation and its rarity itself make it difficult to diagnose. Here we intend to highlight the importance of key diagnostic investigations such as chest X-rays, echocardiograms and computerized tomography in arriving at the diagnosis. A high degree of suspicion is required to arrange crucial investigations without delay.

Keywords: Scimitar syndrome, respiratory distress, pulmonary hypertension

Introduction

George Cooper, in 1836, first described the Scimitar syndrome. It comprises a collection of congenital cardiac anomalies involving partial anomalous pulmonary venous drainage from the right lung into the inferior vena cava (IVC), varying degrees of right lung hypoplasia, and dextroposition of the heart due to lung hypoplasia, pulmonary sequestration and systemic collaterals from the aorta. Other cardiac anomalies might present, such as atrial septal defect (80%), patent ductus arteriosus (75%), ventricular septal defect (30%) and pul-

monary valve stenosis (20%) [1-2]

Among the congenital cardiac conditions, Scimitar syndrome is rare. Though the documented prevalence is 1-3 per 100000 live births, it may be overlooked as some remain asymptomatic through adulthood. Depending on the presentation, two types of Scimitar syndrome are categorised. Namely, the Infantile form, which presents early in life with tachypnea symptoms, repeated chest infection, heart failure, cyanosis, and failure to thrive and the Adult form, which is usually an incidental finding [2].

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The anomalous vein draining the right lung into the IVC traverses the right lung along the right heart border, creating a radiographic density shaped like a scimitar (a curved Turkish sword). Hence the term scimitar syndrome is derived. However, this classic radiographic feature will not be evident in all cases of scimitar syndrome making the diagnosis difficult, as in our case [3].

As for our thorough literature survey, we could not find any reports which elaborate the diagnosis and management of scimitar syndrome in infants in the Sri Lankan context. Previously, an adult with scimitar syndrome was reported in Sri Lanka, which was diagnosed when she was evaluated for palpitations.

Case presentation

A 2-month-old male infant was transferred from a local hospital for further evaluation after being treated for three separate lower res-

piratory tract infections, first needing intubation as well. The child had failed to thrive and had respiratory distress in between infections as well. This is the first child born to healthy non-consanguineous parents after an uneventful antenatal period. On admission, the child weighed 3.9 Kg (-3SD). He had a respiration rate of 32/minute and a heart rate of 140 bpm. Auscultation revealed bilateral lung rales and a grade 2/3 systolic murmur at the left sternal edge.

Interval chest X-rays showed congested lungs with opacities, mainly in the right middle and lower zones (Figure 1). The Scimitar sign was not clearly evident. Blood investigations revealed normal full blood counts (White Blood Cells – 9 000/ml, Platelet – 460 000/ml, Haemoglobin – 15 g/dl) and normal C-reactive protein (5.8 mg/L).

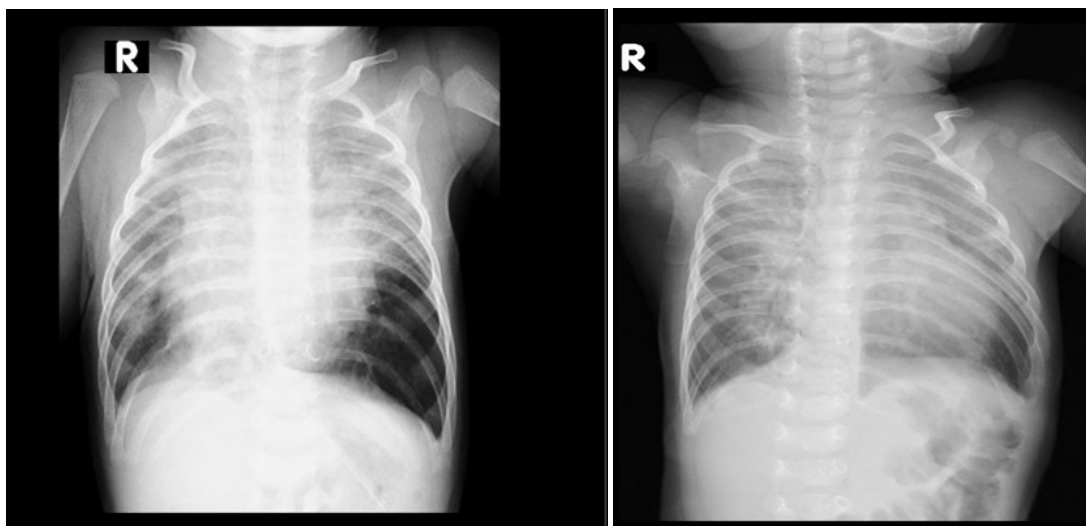


Figure 1: (a) and (b) Chest X-ray PA. Two X-rays were taken 2 weeks apart. Both show the Mesocardiac position of the heart. Figure 1 (a) shows bilateral opacities involving the right upper and middle lobes and left upper lobes. This was taken at 3 weeks of life. The baby was treated for bronchopneumonia. Figure 1 (b) shows a chest X-ray taken 2 weeks after. Bilateral opacities have reduced. However, the Scimitar sign suggestive of the Scimitar vein is not evident.

The echocardiogram showed situs solitus with mesocardia and Right hemi anomalous pulmonary vein draining into IVC. Left pulmonary veins drain into Left Atrium. Moderate-sized ostium secundum Atrial Septal Defect with dilated Right Atrium and Right Ventricle and moderate pulmonary hypertension. Tricuspid Regurgitation peak pressure gradient was recorded at 30 mmHg. The study showed normal ventricles, pulmonary arteries, aortic arch, and

cardiac valves. There was no Patent Ductus Arteriosus.

A contrast CT Angiogram of the chest confirmed the scimitar vein (PARPV) and showed lung intra-parenchymal Arterio-Venous malformation with systemic arterial supply. There was no right lung hypoplasia (Figure 2). Further, cardiac catheterisation showed aorto-pulmonary collaterals with moderate pulmonary hypertension.



Figure 2: Contrast CT Angiogram

The contrast CT scan shows the scimitar vein that traverses through the right lung parenchyma (Red arrow), which enters the intra-hepatic part of the inferior vena cava. It also shows the AV malformation in the right lung. The Mesocardiac position of the heart is also clearly seen.

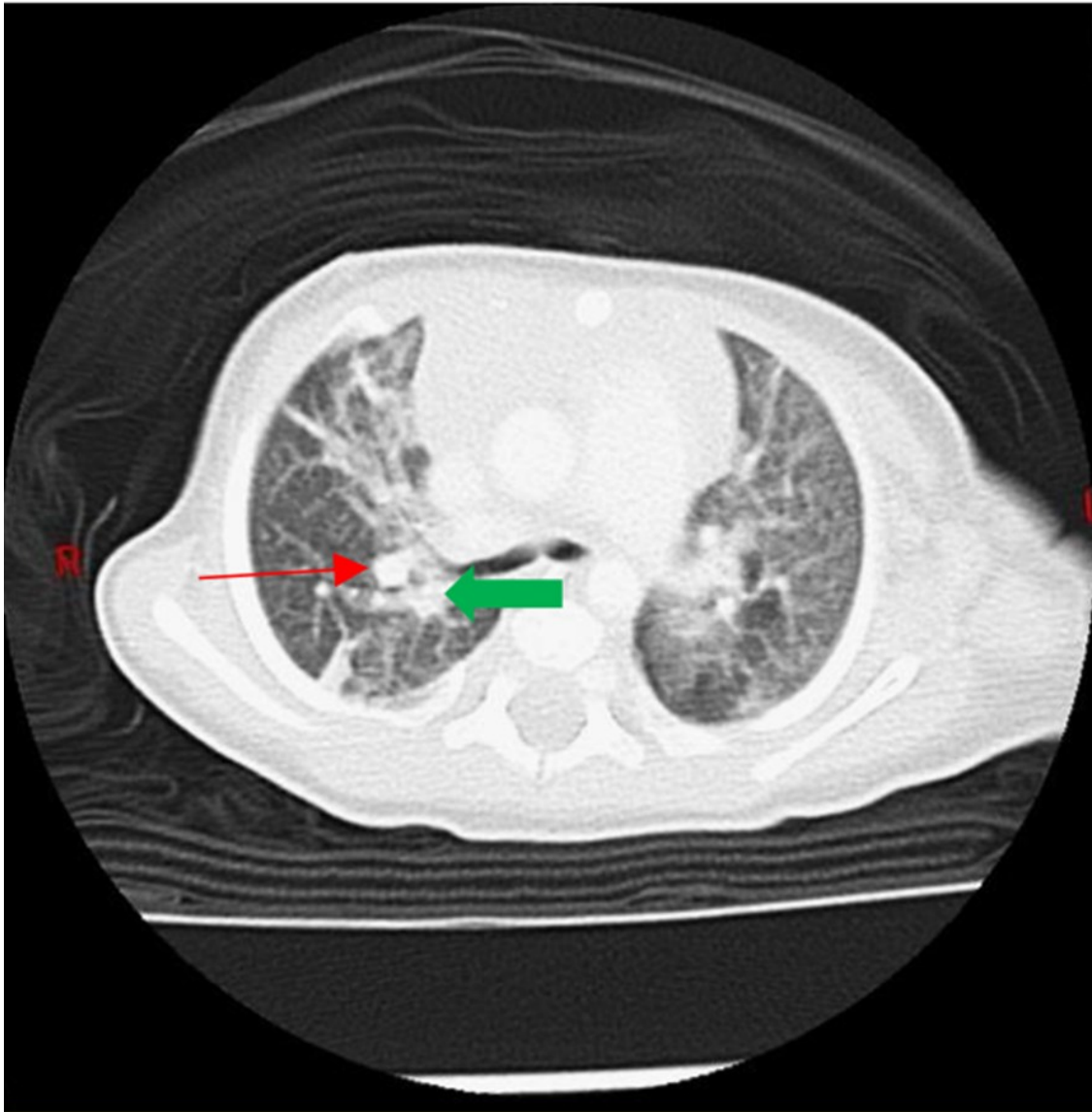


Figure 3: Contrast CT scan of chest – the lung window showing the right side superior pulmonary vein traversing the right lung parenchyma to drain into intrahepatic IVC (Red arrow). The right phrenic artery appears to be dilated and is seen extending upward into the pulmonary parenchyma adjacent to aberrantly draining superior pulmonary vein giving rise to a tuft of vessels (green arrow).

Coil occlusion of aortopulmonary collaterals was done, and follow-up echocardiograms showed resolving pulmonary hypertension. The latest echocardiogram (at 11 months of age) showed only mild pulmonary hypertension. Tricuspid Regurgitation peak pressure gradient dropped to 18 mmHg following the interven-

tion. However, the left to right shunt through the scimitar vein and moderate Ostium Secundum-ASD remain, causing features of heart failure and poor weight gain. He is awaiting re-routing of the anomalous right pulmonary vein into the left atrium and the closure of moderate ASD in due course.

Discussion

Scimitar syndrome represents just 3-5% of all PAPVD. It is an uncommon congenital syndrome that presents with dextroposition, hypoplasia in the right lung, malformation of the right lung, misconnection of the right pulmonary veins to the IVC-right atrial junction, and unusual transmission of the systemic arterial circulation to the lower segments of the right lung. Presentation in infancy may show signs of pulmonary hypertension, heart failure, respiratory distress, or even be asymptomatic in some infants [1-3].

In most case reports we reviewed, the diagnosis was guided by the chest x-ray finding of the scimitar sign. An echocardiogram and CT scan were done in order to confirm the diagnosis. However, in some cases, the classical X-ray finding will not be evident, as in our case. Hence echocardiogram played a major role in arriving at the diagnosis. The absence of the scimitar sign in the X-ray was the main reason for the delayed referral of this child to our centre [4].

There were some case reports that documented other associated physical abnormalities. For example, an Ethiopian report mentions a neonate with scimitar syndrome who had imperforate anus [5]. Our child did not have any associated anomalies.

Most case reports show the presence of dextrocardia and significant right lung hypoplasia. Our child, however, had mesocardia. The presence of mesocardia with the syndrome is less documented in the literature. Most case reports

show right lung sequestration fed by systemic arterial supply [6]. Our child has intraparenchymal arteriovenous malformation with a systemic blood supply, contributing to elevated pulmonary hypertension.

As mentioned, the clinical presentation is a spectrum; some may remain asymptomatic until adulthood. A case report described a 38-year-old adult woman who presented with dyspnoea and fatigue for one week. Her chest x-ray showed dextrocardia and a right opacity. A CT scan of the chest revealed an accessory lobe of the liver extending into the chest cavity through a defect in the posterior right hemidiaphragm. It also showed the right pulmonary vein draining into the IVC, consistent with scimitar syndrome [7].

The management of Scimitar syndrome diagnosed in early infancy depends on the severity of pulmonary over circulation, heart failure, and associated pulmonary hypertension. In symptomatic infants with significant left-to-right shunting, early surgical intervention is often necessary to re-route the anomalous pulmonary vein into the left atrium and close any associated atrial septal defects. Pulmonary hypertension should be addressed preoperatively, sometimes requiring transcatheter interventions such as coil embolization of aortopulmonary collaterals to reduce pulmonary over circulation. Supportive measures, including optimizing nutrition and managing heart failure symptoms with diuretics if needed, are crucial until definitive repair is performed. Postoperative follow-up includes

echocardiographic monitoring to assess pulmonary pressures, right heart function, and residual shunting (6).

Conclusion

Our child had a rare clinical entity called scimitar syndrome. Getting a cardiologist's opinion and performing an echocardiogram was key to arriving at a diagnosis. The CT scan confirmed the cardiologist's suspicion. We shared this case to highlight the importance of advanced imaging techniques in diagnosing rare cardiovascular abnormalities, especially when the preliminary imaging findings are inconclusive.

Abbreviations

PARPV - Partially anomalous right pulmonary vein

IVC – Inferior vena cava

CT – Computer Tomography

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Consent to publish the declaration

Informed written consent was obtained from the child's parents to publish the information and accompanying images about the child's illness.

Data availability statements

The authors' data supporting this study's findings are available upon reasonable request.

Contributions

HP, CL, RG, and SP contributed to the patient's diagnosis and management. HP and SP prepared the manuscript for publication. All au-

thors read and finalised the manuscript

Competing interests

The authors declare that they have no competing interests

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