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Case Report

Pulmonary Hypertension with Atrial Septal Defect in an Infant: A Case Report of Rare Presentation of Scimitar Syndrome?

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ABSTRACT

Background: Scimitar syndrome is characterized by partial or total anomalous pulmonary venous return from the right lung along with pulmonary hypoplasia. Scimitar syndrome is associated with a partial anomalous pulmonary venous connection of the right lung to the inferior vena cava, right lung hypoplasia, heart dextroposition, and anomalous systemic arterial supply to the right lung. Patients are diagnosed either early with severe symptoms (infantile type) including tachypnea, chest infection, heart failure and failure to thrive or late with minimal symptoms (childhood/adult type) as a result of accidental findings. The most common anomalies associated with this syndrome are intracardiac defects with a prevalence of about 40%. Here,

Case report: we present a case of a male infant with respiratory distress and manifestation of large ASD2 and pulmonary hypertension, who was diagnosed with scimitar syndrome after more evaluation.

Conclusion: This diagnosis should be considered when ASD2 with PH are diagnosed in infantile period.

Keywords: Congenital anomaly, Pulmonary hypertension, Scimitar syndrome

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Introduction

Scimitar syndrome was first reported by Naill in 1960, as the radiographic appearance of the anomalous vein that is manifested as a tubular opacity parallel to the right heart border, which simulates a curved Turkish sword or a Scimitar, and hence, the term Scimitar sign. This anomaly has an occurrence rate of about 1 to 3 per 100,000 live births (1). Since many patients do not show any symptoms, true occurrence may be higher.

Anomalous pulmonary venous drainage (APVD) accounts for 2% of all congenital heart diseases. Scimitar syndrome is characterized by a partial anomalous pulmonary venous connection of the right lung to the inferior vena cava, right lung hypoplasia, heart dextroposition, and anomalous systemic arterial supply to the right lung (1). Patients are diagnosed either early with severe symptoms (infantile type) including tachypnea, chest infection, heart failure and failure to thrive or late (childhood/adult type) with minimal symptoms as a result of accidental findings (2, 3). Infantile group of patients has high mortality rate and management difficulties, since the symptoms of severe pulmonary hypertension develop soon after birth (3-5). There are two different types of scimitar veins, the first type is simple classic vein that runs from the middle of the right lung to the cardiophrenic angle and the second type is double arched vein in the upper and lower lung zones, with drainage into the left atrium and inferior vena cava (6, 7).

Patients with Scimitar syndrome who are identified early in life usually have associated congenital heart diseases. In the presence of significant left-to-right shunt and pulmonary hypertension, surgical intervention should be considered. In this case report, we present a male infant with clinical manifestation of tachypnea and poor weight gain, who was diagnosed with scimitar syndrome after more evaluation.

Case presentation

A male infant was examined due to poor weight gain and tachypnea after showing no symptoms in the first 18 months of his life. The physical examination showed that he had S2 splitting and holosystolic murmur in the lower left sternal border (LLSB). There was no evidence of hepatomegaly, ascites or a prominent jugular venous pressure (JVP). Respiratory rate was 60 per minute and the right arm blood pressure was 78/45 mm Hg. Pulmonary sounds were normal and clear.

At 18 months of age, echocardiography demonstrated a large atrial septal defect (ASD), right ventricular enlargement (RVE), right atrial enlargement (RAE), severe tricuspid regurgitation (TR) with moderate right-sided pressures, and normal size of main pulmonary artery. The diameter of the visualized right and left pulmonary arteries was normal. At the level of the hepatic vein (HV) and inferior vena cava (IVC), the scimitar vein was imaged. Color flow Doppler showed flow from the scimitar vein to the IVC. Pulse wave Doppler examined the hepatic vein flow. The PWD of the scimitar vein showed rapid monophasic waveform compared to the characteristically low velocity and biphasic waveform of the HV.

The electrocardiography (ECG) revealed RVE and RAE without right bundle branch block (RBBB) without any sign of ischemia.

The chest X-ray (CXR) did not show any silhouette sign on the right border of the heart, and there was no sign of cardiomegaly and increased pulmonary vascular marking (PVM).

CT scan demonstrated levocardia, anomaly of pulmonary venous drainage from right lung into inferior vena cava, and three aortopulmonary collaterals arising from the aorta.

Elective cardiac catheterization was performed. The pulmonary arterial pressure (55/25 mmHg, mean pressure: 45 mmHg) and systemic systolic pressure were normal. A hypoplastic anomaly of the right pulmonary vein connecting to the inferior vena cava was noted on the levophase of the angiogram.

In our case, angiography demonstrated that two of four pulmonary veins drained into the left atrium but lower and middle right pulmonary veins drained into IVC at the lower diaphragm level. The patient was scheduled for the surgery. Using sildenafil improved pulmonary hypertension after successful operation (Figure 1).

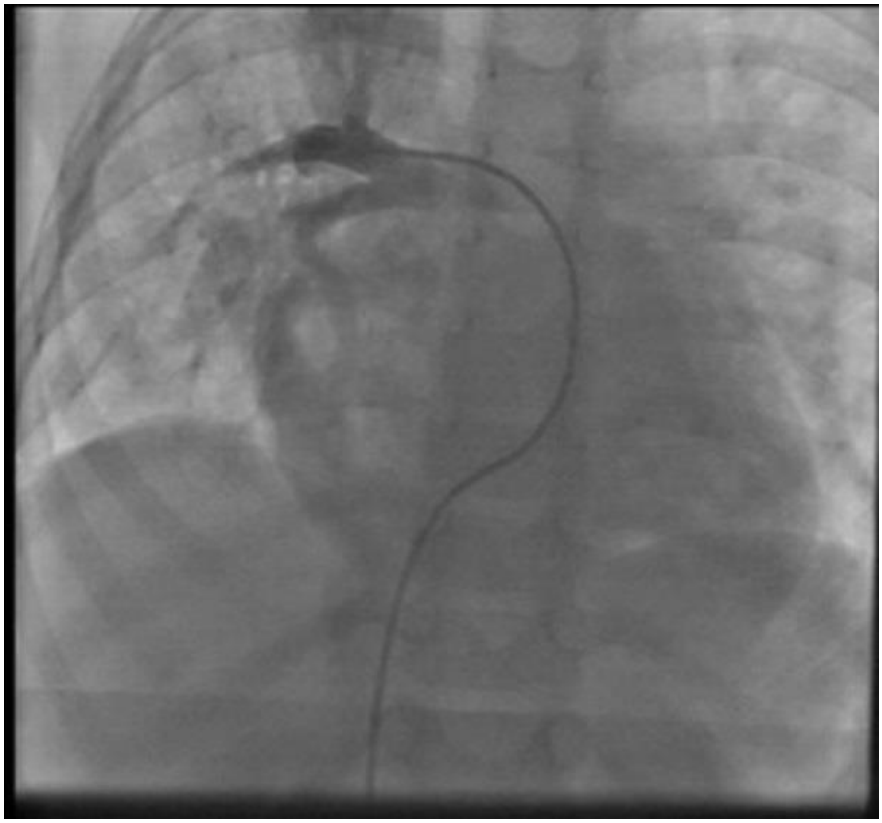

Figure 1. Angiography showing right pulmonary veins drained into the IVC at the lower diaphragm level and the second type is double arched vein in the upper and lower lung zones, with drainage into the left atrium and inferior vena cava, (venous access: 1- inferior vena cava (IVC), 2- right atrial, 3- right ventricular, 4- main pulmonary artery, 5- right pulmonary artery),*showing anomalous pulmonary vein return to IVC.

Discussion

Pulmonary hypertension, as one the best indicators of scimitar syndrome, is often identified by severe symptoms and it shows a poor prognosis during infancy. There are many factors contributing to pulmonary hypertension including large left-to-right shunt via the anomalous pulmonary vein, right lung hypoplasia with reduced pulmonary vascular bed, pulmonary vein stenosis and obstruction, and persistent pulmonary hypertension of the newborn (PPHN) (4, 6, 7).

Pulmonary vasodilators such as Inhaled nitric oxide (iNO), Sildenafil, Bosentan, and Milrinone cause significant improvement in the treatment PPHN.

The previous study showed that the addition of oral Sildenafil to intravenous Milrinone was associated with better therapeutic outcomes in the treatment of PPHN (8).

Hemodynamically, there is an acyanotic left-to-right shunt. The anomalous vein usually drains into either inferior vena cava (most common), right atrium or portal vein. Draining a part or the entire of right lung into the inferior vena cava (IVC) occurs with an anomalous right pulmonary vein, which is the hallmark of scimitar syndrome (6, 7).

It has been reported that 19–31% of patients with scimitar syndrome have associated with congenital cardiac anomalies, and atrial septal defect, with a probability of 70%, is the most common anomaly (6).

Classic findings on physical examination include a shift in heart sounds and cardiac impulse to the right side of thorax and a systolic murmur. Auscultation of the lung is usually normal, although breath sounds may be reduced on the right side of chest (9, 10). The clinical spectrum of scimitar syndrome ranges from severely ill infants to asymptomatic in the youngest children.

Clinical symptomatology of the scimitar syndrome is based on the age at which the syndrome presents. Infants with scimitar syndrome present hemoptysis cyanosis, poor growth, PAH, and often complex cardiac defects, many of which need surgical intervention with a high mortality rate (3).

In older children, it commonly presents with recurrent pulmonary infections and/or exertional dyspnea. Hemoptysis as a presenting symptom is exceptionally rare in patients with scimitar syndrome (5). Interestingly, according to the literature, the youngest patient presenting with hemoptysis was 7-year-old. The possible mechanisms of hemoptysis in scimitar syndrome include rupture of hypertrophied systemic pulmonary anastomosis (5, 11). Pulmunary Artery Hypertension (PAH) is a problem of infancy and it rarely presents in a patient aged 14 years. It has been shown that pulmonary arterial pressure greater than 50 mm Hg is exceedingly rare in patients who present beyond the infancy. Furthermore, it has been demonstrated that PAH in this population may be associated with cardiac anomalies. This could be explained by higher levels of anomalous pulmonary venous drainage, greater extent of hypoplasia of right lung and pulmonary artery, and increased systemic blood flow to the hypoplastic lung (3, 5).

This syndrome is chiefly diagnosed by the presence of characteristic radiological sign (scimitar sign) on conventional chest radiography. However, when scimitar vein is masked by cardiac shadow, diagnosis can be made using one or more modalities such as angiography, CT scan, and echocardiography. The MR technology also shows excellent visualization of vascular anatomy of this complex congenital defect noninvasively (2, 12).

Three-dimensional CT and cardiac-gated MRI are useful for visualizing the anomalous pulmonary vein. They can be particularly helpful in finding an associated horseshoe lung, in which there is a posterior fusion of portions of the right and left lungs behind the heart and before the esophagus and spine. Approximately 80% of infants with horseshoe lung also have scimitar syndrome (13).

Considering the wide clinical spectrum of scimitar syndrome, the medical intervention depends on the severity of presentation and the amount of blood flowing to the IVC from completely or partially anomalous pulmonary veins. When there is a small amount of drainage, therapy may not be required. The use of antibiotics for chest infections, the promotion of good nutrition, oxygen supplementation, and the prescription of sildenafil for pulmonary hypertension could be effective in medication and prognosis. The respiratory symptoms are typically one of the main indications of surgical correction (14, 15). In the presence of significant left-to-right shunt and pulmonary hypertension, surgical intervention should be considered.

In this study, it was observed that advanced pulmonary hypertension medications were useful as the first-line treatment. Surgical approaches to scimitar syndrome vary according to the anatomic and pathologic features presented in each case (16).

Conclusion

Scimitar syndrome is an infrequent disease with diverse presentations and associations, as described in this paper. Thus, diagnosing infantile scimitar syndrome requires meticulous attention and high suspicion of the early referral and management. The association of the syndrome with pulmonary hypertension leads to recurrent and prolonged hospitalization. This condition can be initially suspected from a chest X-ray, but it is typically confirmed via CT angiography. In the scimitar syndrome that presents in infants, high blood flows into the IVC from the anomalous pulmonary veins are negative prognostic factors. This diagnosis should be considered when ASD2 with PH are diagnosed in infantile period.

References

1. Neill CA, Ferencz C, Sabiston DC, Sheldon H. The familial occurrence of hypoplastic right lung with systemic arterial supply and venous drainage "scimitar syndrome". Bulletin of the Johns Hopkins Hospital. 1960;107:1-21.
2. Tamimi OR, Al-Ghaihab M, Al Akhfash AA. Transcatheter closure of patent ductus arteriosus in infantile form of Scimitar syndrome with a type II Amplatzer Ductal Occluder. Journal of the Saudi Heart Association. 2010;22(2):65-7.
3. Uthaman B, Abushaban L, Al-Qbandi M, Rathinasamy J. The impact of interruption of anomalous systemic arterial supply on scimitar syndrome presenting during infancy. Catheterization and cardiovascular interventions : official journal of the Society for Cardiac Angiography & Interventions. 2008;71(5):671-8.
4. Juraszek AL, Cohn H, Van Praagh R, Van Praagh S. Isolated left-sided scimitar vein connecting all left pulmonary veins to the right

inferior vena cava. Pediatric cardiology. 2005;26(6):846-7.

5. Reddy R, Shah R, Thorpe JA, Gibbs J. Scimitar syndrome: a rare cause of haemoptysis. European journal of cardio-thoracic surgery : official journal of the European Association for Cardio-thoracic Surgery. 2002;22(5):821.

6. Khan A, Ring NJ, Hughes PD. Scimitar syndrome (congenital pulmonary venolobar syndrome). Postgraduate Medical Journal. 2005;81(954):216-.

7. Mordue BC. A case series of five infants with scimitar syndrome. Advances in neonatal care : official journal of the National Association of Neonatal Nurses. 2003;3(3):121-32.

8. Bagheri MM, Ebrahimpour M, Amirzadeh R, Derakhshan R. Comparison of the Efficacy of Milrinone Injection versus Milrinone Plus Oral Sildenafil in Newborns with Pulmonary Arterial Hypertension. Journal of Kerman University of Medical Sciences. 2021;28(1):1-9.

9. Agayev A, Yekeler E. Left-sided scimitar syndrome. Pediatric radiology. 2009;39(2):191.

10. Wang CC, Wu ET, Chen SJ, Lu F, Huang SC, Wang JK, et al. Scimitar syndrome: incidence, treatment, and prognosis. European journal of pediatrics. 2008;167(2):155-60.

11. Argueta-Morales IR, Meador LC, Nykanen DG, DeCampli WM. Infantile form of scimitar syndrome with contralateral pulmonary vein stenosis. Pediatric cardiology. 2010;31(4):550-2.

12. Sinha R, Singh P, Bhatnagar AK, Batra A. Scimitar syndrome: imaging by magnetic resonance angiography and Doppler echocardiography. The Indian journal of chest diseases & allied sciences. 2004;46(4):283-6.

13. Inoue T, Ichihara M, Uchida T, Sakai Y, Hayashi T, Morooka S. Three-dimensional computed tomography showing partial anomalous pulmonary venous connection complicated by the scimitar syndrome. Circulation. 2002;105(5):663.

14. Kahrom M, Kahrom H. Scimitar syndrome and evolution of managements. The Pan African medical journal. 2009;3:20.

15. Vida VL, Speggiorin S, Padalino MA, Crupi G, Marcelletti C, Zannini L, et al. The scimitar syndrome: an Italian multicenter study. The Annals of thoracic surgery. 2009;88(2):440-4.

16. Brown JW, Ruzmetov M, Minnich DJ, Vijay P, Edwards CA, Uhlig PN, et al. Surgical management of scimitar syndrome: an alternative approach. The Journal of thoracic and cardiovascular surgery. 2003;125(2):238-45.