Case Report

Scimitar syndrome – Uncommon variant with common presentation

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ABSTRACT

Scimitar syndrome, or congenital pulmonary venolobar syndrome, is a rare congenital heart defect characterized by the anomalous venous return from the right lung. A number of cases that lack all the features of a typical syndrome have been described as the scimitar variant, but the incidence is rare. These variants may be difficult to diagnose and, if diagnosed, they may not require surgical correction, instead, can be managed through episodic treatment and yearly monitoring for the development of complications. Herein, we report the case of a 9-month-old male baby with an uncommon variant of scimitar syndrome where the aortopulmonary collateral maintaining the blood supply of the right lung aroused from the right subclavian artery and descending thoracic aorta instead of inferior vena cava. Due to this, the management completely changed from catheterization of the heart to just follow-up monitoring of ventricular function and development of severe pulmonary hypertension.

Key words: Congenital heart defect, Pulmonary hypertension, Scimitar syndrome, Scimitar variant.

Scimitar syndrome is a complex association of cardiovascular and bronchopulmonary abnormalities, with the main feature of partial or total anomalous pulmonary venous drainage of the right lung to the systemic circulation [1]. It is characterized by partial anomalous pulmonary venous drainage of the right lung causing left-to-right shunt along with the right lung hypoplasia, dextraposition of the heart [2], and pulmonary hypertension due to aortopulmonary collaterals [3]. When visualized on imaging, it resembles a backsword with a curved blade, also known as scimitar in Middle East [4]. In infants, it can present as recurrent lung infection and can develop complications such as pulmonary arterial hypertension and congestive cardiac failure [5], while in adults, it usually remains asymptomatic [6]. It can be associated with other congenital heart defects such as single ventricular defect causing congestive cardiac failure [7] or atrial septal defects [8]. The surgical treatment includes right-sided pneumonectomy [9] or, a tunnel connecting the scimitar vein, and the left atrium can be made by Dacron graft through intra-atrial baffle technique [10].

CASE REPORT

A 9-month-old male baby presented to the pediatric outpatient clinic with complaints of fever with cough, cold, and decreased oral intake for 5 days. His vitals demonstrated low-grade fever of 98.2°F with a pulse rate of 110/min, and respiratory rate of 36/min. On physical examination, he looked pale and crepitations were heard over his chest. Therefore, he was admitted and commenced on IV antibiotics and nebulization.

The laboratory findings showed anemia (6.70 g/dl), leukocytosis with counts of 13,200/mm^3, and elevated C-reactive protein (CRP) levels of 23.40 mg/dl. The chest X-ray showed diffuse changes of bronchitis. His further laboratory investigations are mentioned in Table 1.

His clinical condition improved over a period of time and there was a decrease in the count and CRP levels. His oral intake also improved gradually and thus was discharged after 3 days with the prescription of oral antibiotics. Following this, he had four bouts of respiratory tract infections with similar complaints and was treated on an outpatient basis.

He underwent a 32 slice computed tomography (CT) scan with IV contrast which demonstrated non-visualization of the right main pulmonary artery and smaller right lung. The 2D echocardiography confirmed the hypoplastic right pulmonary artery with a small left-to-right patent ductus arteriosus shunt. The aortopulmonary collaterals were noted supplying to the right lung in the absence of the right pulmonary artery. These findings were in accordance with the scimitar syndrome variant with preferential blood flow to the left lung causing mild pulmonary artery hypertension.

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Table 1: Laboratory investigations of the patient

Investigations	Visit-1	Visit-2	Visit-3
Hemogram	Low Hb – 7.4 g/dl (13.5–17.0 g/dl) Low total RBC – 3.94 mil/mm^3 (4.6–6.2 mil/mm^3) Low PCV – 25.6% (40–54%) Low MCH – 18.78 pg (27–31 pg) Low MCV – 64.98 fL (80–96 fL) Increased RDW –15.40% (10–15%) High total WBC – 12,300/mm^3 (4000–11,000/mm^3) High platelet count – 4.87 lac/mm^3 (1.5–4 lac/mm^3) Increased C-reactive protein – 10.41 mg/L (00–06 mg/L)	Low Hb – 11.7 g/dl (13.5–17.0 g/dl) Low total RBC – 4.38 mil/mm^3 (4.6–6.2 mil/mm^3) Low PCV – 36.30% (40–54%) Low MCH – 26.71pg (27–31 pg) Lymphocytose – 54% (20–40%)	
Chest X-ray	Diffuse changes of bronchitis in both lungs	Ill-defined consolidation in the right lower zone near CP angle and lower upper zone	Diffuse bronchitis changes with faint consolidation in both lungs
Echocardiogram			Mesocardia Mild dilated right atrium and right ventricle Predominant left-sided pulmonary venous drainage

Hb: Hemoglobin, MCV: Mean corpuscular volume, MCH: Mean corpuscular hemoglobin, RBC: Red blood cell, WBC: White blood cell, PCV: Packed cell volume, RDW: Red cell distribution width, CRP: C-reactive protein, CP angle: Costophrenic angle

He was referred to a pediatric cardiologist for further evaluation and management. Consequently, serial reassessment with 2D echocardiography was advised with an oral combination of furosemide and spironolactone for the reduction of pulmonary arterial hypertension. His progression was complicated by the development of bronchopneumonia for which, he was admitted again. On admission, he appeared ill with a fever of 101.9°F, pulse rate of 109/min, and respiratory rate of 39/min. The chest X-ray was significant for ill-defined consolidation patch in the right lower zone.

He was commenced on IV antibiotics and nebulization for the same. Following his discharge after 3 days, he was referred to a higher center for further evaluation. A 3D construction of heart was done using 128 slice CT scan which confirmed the previous findings of scimitar syndrome. As the patient had a good biventricular function and no signs of cardiac failure, his further management included regular monitoring for the development of pulmonary hypertension and cardiac failure.

DISCUSSION

Scimitar syndrome is characterized by partial or complete venous communication of the right lung into the inferior vena cava. Some rare variants such as scimitar veins draining to the left atrium [1,2] and bilateral anomalous pulmonary venous connection have also been documented [3]. It can be associated with other congenital heart defects such as single ventricular defect causing congestive cardiac failure [4]. The atrial septal defects are also noted to coexist with the scimitar syndrome [5].

The chief pathophysiological mechanism includes the development of left-to-right shunt due to the abnormal return of one or more right pulmonary vein(s), which depends on the pulmonary lobe or segments from which the anomalous vein originates, number of pulmonary veins involved, and relative resistance of draining pulmonary beds [6]. This shunt eventually induces volume overload on the right-sided heart, thus causing heart failure [7]. The right lung and right pulmonary hypoplasia contribute to modulate the left-to-right shunt. Thus, the patient with severe hypoplasia develops a consequently little volume overload and, therefore, has no symptoms. However, sometimes significant hypoplasia of the lung may lead to the development of hypoxemia. In this case, the patient may present with recurrent pneumonia or recurrent lower lobe infections [8], our patient also had the same clinical picture. All of the above-mentioned abnormalities may cause pulmonary hypertension in the long term and this becomes the main reason for poor outcomes of the disease [9].

The diagnosis of scimitar syndrome can be established through various techniques. Clinical examination can support the diagnosis in the case of symptomatic patients. The electrocardiogram may show some non-specific signs like prominent R wave in leads V1 to V3 due to dextroposition of the heart. Hypoplastic right lung or the right hemithorax mediastinal shift due to dextroposition of the heart and blunt costophrenic angle can be seen on chest X-ray. Two-dimensional echocardiography and CT of the chest can delineate lung anatomy and the scimitar vein, as well as, the systemic blood supply to the right lung. Lung scintigraphy can be helpful to assess the right lung perfusion with technetium-99. For three-dimensional reconstruction of the heart, magnetic resonance imaging (MRI) would be helpful. MRI is also helpful as it provides optimal anatomical visualization of anomalous draining vessels and it does not use ionizing radiation as well (Fig. 1).

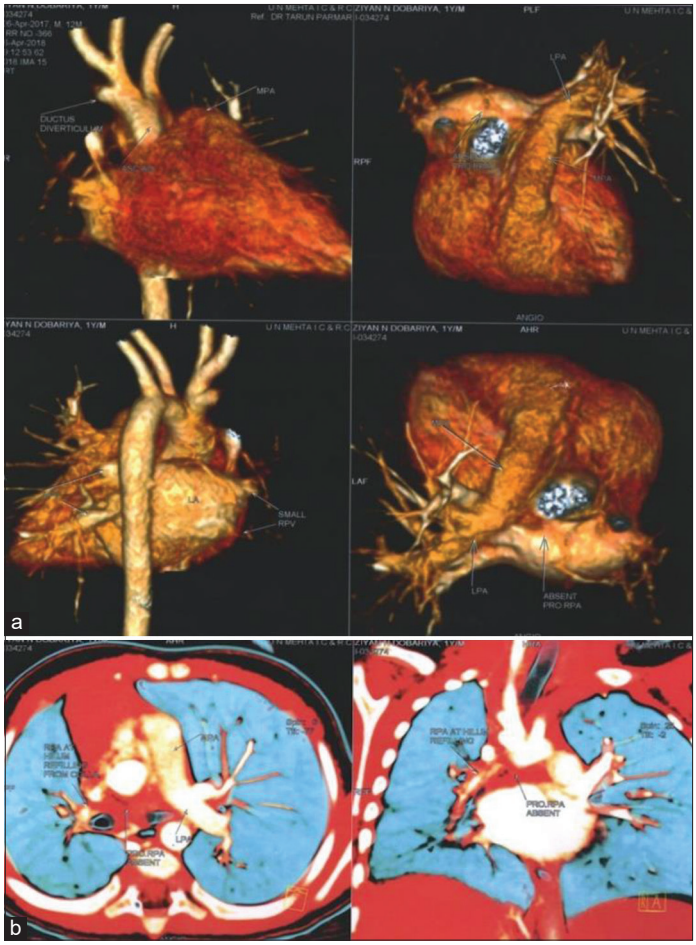

Figure 1: (a) The 3D reconstruction of the heart and its vasculature using a multiscan cardiac computed tomography scan demonstrating hypoplastic right pulmonary trunk, while the left pulmonary trunk is visualized normally; (b) The pulmonary vasculature with the help of contrast demonstrating the clear absence of the right pulmonary artery along with patent left pulmonary artery

The treatment modalities include right-sided pneumonectomy or creating a tunnel connecting the scimitar vein and the left atrium made by Dacron graft through intra-atrial baffle technique [10]. Embolization of the aortopulmonary collaterals can also be done by detachable coils [11]. During our literature search, we came across other case reports of scimitar syndrome associated with other congenital defects such as VACTERL, H-type tracheoesophageal fistula [5,12], and imperforated anus [13]. We also came across case reports showing features of failure to thrive due to recurrent chest infections, leading to immediate repair of the defect. In contrast, in our case, the ventricular function was completely normal. Thereby, follow-up was the mainstay of the treatment with regular 2D ECHO checks.

CONCLUSION

Scimitar syndrome is a rare congenital cardiac disorder that can present with varied anatomy. Thus, it becomes paramount to understand the anatomy of the different variants of the syndrome. As in this case, we have vasculature arising from the right subclavian artery as well as the descending thoracic aorta.

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