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Two Cases Report of Scimitar Syndrome: The Classical one with Subaortic Membrane and the Scimitar Variant

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Scimitar syndrome is characterized by an anomalous pulmonary vein draining into the inferior vena cava, visible roentgenographically as a crescentic shadow of vascular density along the right border of the cardiac silhouette and dextroposition of the heart. We are reporting two cases of Scimitar sydrome; one being classical associated with subaortic membrane and the other a Scimitar variant. The clinical and anatomical spectrum for each case is described.

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Scimitar syndrome is a rare but well described constellation of cardio-pulmonary anomalies: anomalous pulmonary venous connection of the right lung, hypoplsia and malformation of the right pulmonary artery and bronchial trees, and anomalous systemic arterial blood supply to the right lung arising from the abdominal aorta or its branches[1]. The partial or total anomalous venous connection of the right lung drains into the inferior vena cava or azygous system. Often this anomalous vessel is visible on a chest radiograph as a curvilinear shadow just above the right diaphragm and resembles a Turkish sword or Scimitar.

The term Scimitar syndrome was first used by Neil et al to describe this disorder[2]. Additional cardiac anomalies are common, such as ventricular septal defect, patent ductus arteriosus, coarctation of the aorta and the tetralogy of Fallot[3]. Other variations in pulmonary venous connection to the left atrium have been described in the Scimitar syndrome[4-8]. We herein report two cases of Scimitar syndrome.

CASE 1

A 3.5 year old girl discovered to have heart murmur and dextrocardia at the age of two and a half years, when she presented with recurrent respiratory infections. She was a product of full term normal vaginal delivery, and family history revealed no similar condition. Her weight was 12.5 Kg (<5% centile) and her height was (<5% centile).

The cardiac examination revealed ejection systolic murmur grade III/VI and an early diastolic murmur grade II/VI over the third intercostal space at the left sternal border. Air entry was decreased on the right lower lung field with increased dullness, the rest of the

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clinical examination was normal. A chest radiograph demonstrated hypoplasia of the right lung, shift of the mediastinal structures to the right and curvilinear densities in the right chest (Scimitar sign) (Fig 1).

Figure 1. CXR: showed dextrocardia and Scimitar sign

Figure 2. 2D-Echo: parasternal long axis view showed subaortic membrane

A 2D-Echocardiogram showed subaortic membrane (Fig 2) with maximum pressure gradient of 50mmHg and grade II aortic regurgitation associated with right atrial and right ventricular enlargement. At cardiac catheterization an oxygen saturation and pressure study showed a step up of oxygen at inferior vena cava and right atrium associated with subaortic membrane with a pressure gradient across the left ventricular outflow tract of 60mmHg and mild pulmonary hypertension. A main pulmonary angiogram with levophase revealed tortuous right upper and lower pulmonary veins which were draining to the inferior vena cava (Fig 3), a left ventricular angiogram showed subaortic membrane.

Figure 3. Main pulmonary artery angiogram showed tortuous right pulmonary veins drained to the IVC

CASE 2

A 12 year old boy was evaluated because of effort intolerance and recurrent chest infections mainly on the right side of one year duration, prior to this presentation he was

completely asymtomatic. He was a product of full term normal vaginal delivery. Family history revealed no similar condition. His weight was 44 Kg (70% centile) and height 141cm (30% centile). The cardiac examination was normal. Air entry was decreased on the right lower lung field with increased dullness. No other abnormal clinical findings were detected.

A chest radiograph demonstrated hypoplasia of the right lung, shift of the mediastinal structures to the right and curvilinear densities in the right chest (Scimitar sign). An electrocardiogram showed normal axis and no atrial or ventricular enlargement. A 2D-Echocardiogram was normal, (the sizes of the right atrium, right ventricle, main pulmonary artery and inferior vena cava were normal).

At cardiac catheterization; oxygen saturations and pressures were normal without intracardiac shunt. A pulmonary angiogram revealed right pulmonary artery hypoplasia, whereas on the levophase the right superior pulmonary vein was enlarged , had tortuous course and was draining into the left atrium. The right inferior pulmonary vein was small, hypoplastic and was draining also into the left atrium (Fig 4).

Figure 4. Levophase showed dilated and tortuous right superior pulmonary vein, and the small hypoplastic right inferior pulmonary vein both are draining to the left atrium

DISCUSSION

The classic definition of the Scimitar syndrome is a triad of hypoplasia of the right lung with anomalous venous drainage and a systemic arterial supply of a variable degree[8]. The typical Scimitar sign was present in 57 of the 67 cases reviewed by Kiely et al[9]. The first case demonstrates classical Scimitar syndrome associated with subaortic membrane a finding which is rarely reported, whereas the second case demonstrates that rarely a false positive Scimitar syndrome may be present with dextroposition of the heart, hypoplasia of the right lung and the Scimitar sign caused by a tortous abnormal pulmonary vein with normal drainage to the left atrium. Similar cases have been rarely reported in literature[4-8].

Other variations in pulmonary venous connection to the left atrium have been described in the Scimitar syndrome. Takeda et al reported 2 cases; in one case the Scimitar vein entered both the inferior vena cava and the left atrium without any intracardiac shunt, in

the other case the Scimitar vein showed a meandering course and then drained into the left atrium[6] and surgical intervention was not required.

The Scimitar sign does require a differential diagnosis because it has been reported in other conditions, some of which are similar to our case[7], these include anomalous intrapulmonary venous connection to superior vena cava left atrium[6] and surgical intervention was not required. These include anomalous intrapulmonary venous connection to superior vena cava[10], anomalous inferior vena cava with normal pulmonary venous drainage[5], obstruction of one of the major pulmonary vein and development of a distended intrapulmonary collateral may produce a Scimitar shadow[10].

The etiology of Scimitar syndrome is unclear but is thought to represent a fundamental abnormality in pulmonary development, mainly involving the right lung, although the left lung can also have abnormal structure[11]. The prognosis of Scimitar syndrome depends on the age of detection. Patients in whom the diagnosis was made during the first year of life had more severe symptoms, higher rate of heart failure and pulmonary hypertension than did the patients in whom the diagnosis was made after the age of one year[12].

CONCLUSION

In conclusion we are reporting two cases of Scimitar syndrome, the first being a classical form associated with subaortic membrane a finding which has been reported rarely and the second rare case of Scimitar variant with positive Scimitar sign, right lung hypoplasia, dextroposition of the heart and malformed right pulmonary venous drainage to the left atrium and in whom surgical intervention was not required.

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