Case 1596

Scimitar syndrome
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Section: Paediatric radiology
Case Type: Clinical Cases
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Patient: 6 years, male

Clinical History:

History of recurrent chest infections.

Imaging Findings:

The patient presented with a history of recurrent chest infections. A PA chest radiograph was taken.

Discussion:

The scimitar syndrome (also known as the congenital pulmonary venolobar syndrome or the hypogenetic lung syndrome) is a rare, complex, congenital disorder affecting the right lung, which was first described by Cooper and Chassnatin in 1836. The incidence is estimated to be 1-3 per 100,000 births, however the true incidence is unknown because the syndrome may remain undetected in asymptomatic patients.

The scimitar syndrome represents a spectrum of abnormalities in which there is hypoplasia or aplasia of one or more lobes of the right lung. The name was introduced by Halas et al. in 1956 and it is derived from the characteristic shape of a large, anomalous vein resembling a Turkish sword, that drains all or part of the right lung. The vein curves inferiorly adjacent to the right heart border before its termination into a systemic vein. The scimitar vein is therefore a form of partial anomalous pulmonary venous drainage (PAPVD). Most commonly the scimitar vein drains infradiaphragmatically into the IVC, but the course of the drainage is variable; drainage into the right atrium, hepatic veins, portal vein or azygous vein has also been reported. More than one anomalous vein may be present.

In addition to PAPVD of the hypogenetic right lung, many other anomalies of the thorax and its vasculature have been described in the scimitar syndrome. Abnormal lobation and bronchial distribution in the right lung, a hyparterial right main bronchus, absence or hypoplasia of the right main pulmonary artery and systemic arterial supply to the hypoplastic right lung are commonly found. Tracheobronchial anomalies including tracheal stenosis and diverticula are recognised. Bronchiectasis is found in approximately 5% of patients. Dextrocardia is not uncommon. Congenital heart disease is present in up to 25% of patients. Typically there is a sinus venosus or secundum type ASD, but VSDs, PDAs, tetralogy of Fallot, double outlet right ventricle and truncus arteriosus have all been reported.

Patients with the scimitar syndrome may be asymptomatic, with the diagnosis being made incidentally on a chest radiograph. Those with symptoms may present with problems due to the associated malformations or to the PAPVD itself. Presentation in infancy is rare but can occur in those infants with concomitant congenital heart disease. Less severe manifestations tend to occur in patients diagnosed in late childhood or adulthood, when the predominant symptoms are of recurrent respiratory tract infections. The PA chest radiograph typically demonstrates a small right hemithorax with ipsilateral shift of the heart and mediastinal structures. The scimitar vein is seen as a soft tissue
density curving adjacent to the right heart border. The right pulmonary artery may be small or absent. All patients being considered for operative repair should undergo some form of angiography. CT or MR angiography may be acceptable to paediatric surgeons as an alternative to conventional arteriography. The abnormal arterial supply to the hypoplastic right lung must be confirmed; this prevents transection of an aberrant artery at surgery. Ventriculography allows visualisation of the abnormal intra- and extracardiac anatomy, and measurement of the left-to-right shunt. The shunt fraction is generally less than 50%.

The management of patients with scimitar syndrome depends on the severity of the patient's symptoms. Asymptomatic patients or patients with mild symptoms do not require surgical intervention. In patients with greater than 2:1 shunts, re-implantation of the anomalous vein into the left atrium may be performed.

**Differential Diagnosis List:** Scimitar syndrome

**Final Diagnosis:** Scimitar syndrome

**References:**


Figure 1

Description: Penetrated PA chest radiograph showing a small right hemithorax, with ipsilateral shift of the heart and mediastinal structures. There is a curved, soft tissue density adjacent to the right heart border (the "scimitar" vein). There is compensatory hyperinflation of the left lung. Origin: