

Scimitar Syndrome

A 20-day-old female child born at term to a primigravida by normal vaginal hospital delivery was admitted in neonatal intensive care unit with respiratory distress. Immediate postnatal period was uneventful. The baby had severe tachypnea, tachycardia and chest retractions. There was no cyanosis and the heart sounds were audible on the left side of the chest with grade II systolic murmur in second left parasternal area. There was no evidence of any external congenital anomaly. Investigations revealed normal hemogram, X-ray chest showed density in the right paraspinal region along with medial inferior lung suggestive of sequestration of right lower lobe (*Fig. 1*), no cardiomegaly or dextroposed heart. Colour Doppler echocardiography revealed ostium secundum ASD with anomalous pulmonary venous drainage. CT scan of chest and abdomen with angiography showed complex cardiac anomaly with aorta draining into the right lung. The consolidated right lower lobe represented an area of sequestration. The patient underwent coil repair of collateral to relieve pulmonary hypertension with clinical improvement of symptoms.

The Scimitar syndrome is a rare condition of cardiopulmonary anomalies(1) accounting for 0.5-1% of congenital heart disease. The incidence of the associated congenital cardiovascular abnormalities is 36% in pediatric age group and is highest (75%) among the neonates and include ASD, VSD, coarctation of aortic arch and abnormal relationship of the pulmonary arteries and bronchi(2). The age of presentation is variable.

In a series of 32 patients over a period of 20 years, the median age at diagnosis was 7 months(3). Severe respiratory insufficiency is always present in symptomatic cases in early age group as Scimitar syndrome results in pulmonary hypertension, heart failure and right lung infection. In older children and adults, the diagnosis of Scimitar syndrome is often made incidentally who undergo chest radiography for diverse reasons. Recurrent respiratory infections and heart murmur may be the mode of presentation in them. Children who are diagnosed with Scimitar syndrome after infancy have fewer associated defects and less pulmonary hypertension.

Treatment for symptomatic Scimitar syndrome consists of surgical repair. Repair of the anomalous venous return and ligation



Fig. 1. Sequestration of right lower lobe.

of colloaterals is generally recommended, although right pneumonectomy also provides similar results(4).

Our patient responded for 6 weeks but then deteriorated and therefore was referred to cardio-thoracic center in Mumbai for further surgical management.

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Acute Transverse Myelitis Following Hepatitis E Virus Infection

A 12-year-old girl presented with sudden onset weakness of both lower limbs for one day, associated with loss of bowel and bladder sensation, leading to overflow incontinence of urine. On examination the child was afebrile and vitals were stable. There was no pallor, icterus, clubbing, lymphadenopathy or edema. Spine was normal. The child was conscious and oriented. There was no cranial nerve involvement. At presentation, muscle tone of all the four limbs was reduced. But in the next 2 days muscle tone normalized. Power at the shoulder joints was 4/5 for all movements. Similarly for elbows it was 4/5 and for wrists and fingers 3/5. Power of all muscle groups of the lower limbs was 1/5. All deep tendon

reflexes turned brisk and abdominal reflex was not elicitable. Plantar reflex was extensor on both sides. Touch, pain, temperature and vibration sensations were decreased all through, from lower limbs till neck. Position sense could not be tested precisely. There were no cerebellar signs or signs of meningeal irritation. Cerebrospinal fluid microscopy and biochemistry were normal.

The child had a history of low-grade fever, anorexia and jaundice (suggestive of viral hepatitis) starting twenty days prior to admission. These symptoms had already resolved before the onset of weakness. Serology was positive for hepatitis E virus antibody (IgM anti HEV). It was negative for surface antigen for hepatitis B (HBsAg) and antibodies for hepatitis A (IgM anti HAV) and hepatitis C (anti HCV). IgM antibodies for measles, rubella and herpes simplex were also negative. Magnetic resonance imaging (MRI)