



PULMONARY VENOBAR SYNDROME

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- Presenting complaint: 10 yrs old girl with recurrent episodes of lower respiratory tract infection from infancy.



- Born out of LSCS, NCM.
- Birth weight 2.7 kg
- Neonatal period uneventful
- Hospitalised for pneumonia at 9 months of age.
- Anti TB treatment received at 2 ½ years at pvt hospital.
- H/O wheeze + , Rx with inhalers.

- Child was also admitted for recurrent lower respiratory tract infection and given iv antibiotics.



CLINICAL EXAMINATION

- Vitals-normal
- Anthropometry- weight-22 kg (-2 to -3 z score)
height-130 cm(-1 to -2 z score)
BMI- 13.01(-2 to -3 z score)
- General examination- N
- Systemic examination-
 - Respiratory system:air entry decreased rt.side.
no added sounds.
 - CVS-S1S2, no murmur.
 - Other systems - normal



INVESTIGATION

Chest X ray

Small lung on right side

Mediastinal shift to the right side

The right heart border blurred.

Anomalous draining vein seen as a tubular structure paralleling the right heart border in the shape of a Turkish sword (“Scimitar”).

ECG- Normal

ECHO- suggestive of PAPVC.



- The scimitar vein connected to the inferior vena cava.
- The left-sided pulmonary venous drainage was normal.
- Hypoplasitic right lung was identified
- No other anomalies in arterial supply to the right lung or in bronchial arteries.



128 SLICE CT PULMONARY ANGIOGRAPHY

- Abnormal pulmonary venous drainage from the right lung via a large straight vein that drained infra-diaphragmatically at the IVC
- A partial obstruction of the anomalous vein noted at its junction with the IVC.



DIAGNOSIS

PULMONARY VENOBAR SYNDROME



MANAGEMENT

- Corrective surgery done
- Scimitar vein is anastomosed directly to the left atrium through a right thoracotomy.

Patient advised -Follow up after 6 months.



DISCUSSION

- Pulmonary Venolobar Syndrome / Scimitar Syndrome.
 - Halasz's syndrome
 - mirror-image lung syndrome
 - hypogenetic lung syndrome
 - epibronchial right pulmonary artery syndrome
 - venacava bronchovascular syndrome.
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- INCIDENCE- 0.5 TO 1% of chd.
 - Female preponderance.



- Pulmonary hypoplasia and partial anomalous pulmonary venous return (PAPVR).
- Right side.
- Haemodynamically, there is an acyanotic left to right shunt.
- The anomalous vein usually drains into
 - IVC : most common
 - right atrium
 - portal vein
- The lung is frequently perfused by the aorta, but the bronchial tree is still connected and thus the lung is not sequestered.



Associations

- congenital heart disease (e.g. ASD, VSD, tetralogy of Fallot, PDA)
- ipsilateral diaphragmatic anomalies (e.g. accessory diaphragm, diaphragmatic hernia)
- localised bronchiectasis
- horseshoe lung
- vertebral anomalies e.g. hemivertebrae
- genitourinary tract abnormalities



CLINICAL FEATURES

Neonatal period :

- Respiratory and/or cardiac failure .
- This is most commonly caused by pulmonary hypertension due to cardiac and/or right lung anomalies.
- Heart failure may also be caused because of a large arterial supply from the abdominal aorta to a sequestered lobe.



Infancy and childhood:

- Recurrent respiratory infections, usually affecting the right lower lobe that often has an abnormal blood supply and venous drainage.
- Severity and frequency of infections is related to the degree of pulmonary hypoplasia.
- Affected individuals may also present with haemoptysis due to pulmonary hypertension .

At any stage in life:

- As an incidental finding e.g. due to the detection of a murmur or due to the evident CXR abnormalities .



Pulmonary hypertension as a marker for Scimitar syndrome:

- Large left to right shunt via the anomalous pulmonary vein.
- Left to right shunt from the systemic arterial supply to the right lung .
- Right lung hypoplasia with reduction of the pulmonary vascular bed.
- Pulmonary vein stenosis and obstruction.
- Other congenital cardiac malformations.
- Persistent pulmonary hypertension of the newborn



INVESTIGATION

Chest X-ray:

- The abnormal venous return is the main component of Scimitar syndrome, and gives a characteristic abnormal radiographic shadow descending along the right cardiac border, which resembles a curved Turkish sword (i.e., Scimitar) .
- Dextroposition of the heart along with varying degrees of opacity of the right hemithorax .
- A small, opaque hemithorax generally implies volume loss.



Echocardiography :

- Delineate both the Scimitar vein as well as any systemic arterial supply to the right lung .
- Additional cardiovascular anomalies have been noted in 75% of infants with severe symptoms.

Fetal echocardiography:

- Permits prenatal diagnosis in which spectral and color Doppler provides clues to the presence of an obstructed pulmonary venous pathway .
- Visualization of a confluence behind the right atrium and a vertical vein are the most consistent Echo clues .



- The flow pattern in Scimitar vein is monophasic extending throughout the cardiac cycle with no reverse flow at atrial contraction .
- Three-dimensional computed tomography (CT) and cardiac-gated magnetic resonance imaging (MRI) are useful in visualizing the anomalous pulmonary vein.
- Helpful in detecting an associated horseshoe lung



- Cardiac catheterization and angiography : gold standard to know the anatomy.
- Oxygen saturations in the IVC and RA may be increased . The RPA is almost always hypoplastic, atretic, or otherwise abnormal in those infants with pulmonary hypertension .
- An aortogram should also be performed to visualize the presence or absence of an anomalous systemic artery entering the right lower lobe.



MANAGEMENT

- In neonates and young infants : a trial of medical therapy as the initial approach is reasonable to allow an increase in size before repair of the defect.
- However the presence of pulmonary hypertension or lack of response to medical therapy demands prompt surgical treatment.



INDICATIONS FOR SURGERY

- Symptomatic patients
- Associated cardiovascular anomalies
- Left to right shunt $>2:1$



SURGICAL OPTIONS

- Redirecting the abnormal vein to left atrium
- The classic operation-Baffle repair.
- Scimitar vein is anastomosed directly to the left atrium through a right thoracotomy.



- Ligation or embolisation of artery supplying sequestered pulmonary lobe
- Pneumonectomy
- PROGNOSIS
- Good in the absence of associated anomalies
- Dependent on the presence of associated congenital heart disease and its severity.



THANK YOU

