Name of Disease:

Pulmonary Lymphangiectasia (PL)

Clinician:

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Clinical details:

Definition
Pulmonary lymphangiectasia is a rare disease characterised by dilatation of the normal lung lymphatics. These channels drain lymph from both the pleural spaces lining the lung and the pulmonary interstitium into the venous system.

Clinical Presentations
The clinical symptoms of PL can present antenatally (as a cause of nonimmune hydrops foetalis), at birth (as a cause of severe respiratory distress) or in the post natal period with persistent tachypnoea, recurrent cough and wheeze.
The overall incidence of PL is unknown but rare. Although most cases are sporadic there are described cases of affected siblings.

Causes
Dilated lymphatics can be primary or secondary to a wide range of predisposing conditions. The primary group is divided into 3 broad categories: i) recognised syndromal conditions which feature generalised abnormalities of the lymphatic syndrome such as Noonan syndrome, Down syndrome, Turner syndrome etc; ii) a generalised lymphangiectasisa with intestinal involvement and oedema often with hemihypertrophy and iii) pulmonary lymphangiectasia (PL) - the condition described here - which is an isolated lymphangiectatic abnormality confined to the lung and pleural spaces.

Diagnosis and investigations
Stepwise approach to investigation
Chest xray: In the neonatal presentation there may be a unilateral or bilateral ground glass appearance often with pleural effusions.

US chest: Can usefully confirm the presence and architecture of the pleural effusion. An ECHO may confirm the presence of an associated pericardial effusion.

Pleural tap: The effusion is typically chylous if enteral feeding has been established. In addition to triglycerides (> 1.1 mmol/l), there may be a high lymphocyte count (>1,000 cells/microL – 80% lymphocytes))with elevated protein and albumin concentration.

CT scan: There is inhomogeneity of the lung parenchyma on high resolution CT slices with interstitial

densities and effusions.

MRI: This is one of the few lung parenchymal conditions where an MRI scan may be helpful especially if there is a need to limit radiation burden. In milder disease the MRI shows cystic changes in the lung parenchyma and dilatation of the interlobular and interlobar septae.

Lung biopsy: Often a combination of clinical features, pleural tap and xray or CT findings are sufficient to make the diagnosis of PL. Occasionally, an open lung biopsy is useful in distinguishing PL from other forms of interstitial lung diseases although the risk of the procedure, in a very sick neonate, needs to be weighed against the usefulness of its findings. .Histology reveals large, often cystic, endothelial lined channels in the distribution of the normal lymphatics. Inflammatory changes are not present but connective tissue may be prominent.

Scintigraphy: Lymphangiography and lymphoscintigraphy are rarely used in paediatric practice for PL.

Neither bronchoscopy nor infant lung function testing add much to the diagnostic investigations. A BAL may prove useful in the older child to identify superadded infections.

Course

A number of children with PL are stillborn. There is a high mortality risk within the first few hours of birth associated with this condition.
In the absence of significant co-existing abnormalities, there is the potential of a gradual improvement in clinical status with intensive neonatal support.
Survival of the acute illness can lead to a prolonged chronic respiratory course. Common symptoms include recurrent cough and wheeze and repeated infective exacerbations with multiples hospitalisations. A few patients require long term oxygen use.
Gastro-oesophageal reflux and poor growth are common associated features.

Treatment

With improved neonatal care, PL, once considered universally fatal, is however showing an improved outcome although severe forms require aggressive respiratory care.

Treatment is essentially supportive:

- Mechanical ventilation is often required in the neonatal form and high mean airway pressures may be required.
- High frequency oscillation with nitric oxide may be necessary.
- Chest tube drainage of chylous pleural fluid may be prolonged.
- Nutritional therapies with medium chain triglycerides and parenteral nutrition may be necessary for recalcitrant cases of effusion.
- Total protein and immunoglobulin levels may need monitoring and replacing where there is large chylous drainage.

Medical therapies such as antiplasmin and octreotide have been tried in intestinal forms of this condition but have not been universally recognised as useful in PL.
Sclerotherapy of the pleural surfaces may be attempted when pleural drainage is particularly prolonged but is often unsuccessful as there is difficulty in getting the lymph-soaked surfaces to adhere.

Useful references:

Esther CR, Barker PM. Pulmonary lymphangiectasis: diagnosis and clinical course. Pediatric Pulmonoloy 2004; 38:308 - 313

Faul JL et al. J Respir Crit Care Med 2000. Thoracic lymphangiomas. lymphangietasis, lymphangiomatosis and lymphatic dysplasia syndrome. Vol 161:1037 – 1046.

Scott C, Wallis C, Dinwiddie R, Owens C, Coren M. Primary pulmonary lymphangiectasis in a premature infant: resolution following intensive care
Pediatr Pulmonol 2003;35(5):405-6

Web links:

Orphanet: Pulmonary Lymphangiectasia information

A parent derive help and information page for pulmonary lymphangiectasia

BPOLD is funded by the University of Edinburgh Research and Development Fund