

LETTER TO THE EDITOR

Congenital pulmonary lymphangiectasia in patient with pulmonary vein stenosis/atresia

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To the Editor,

An on-term male infant weighing 3500 g was admitted to the hospital due to systemic oxygen desaturation and haemodynamic instability. Apgar score was 8 at 1st minute. A continuous infusion of dopamine was instituted for low-cardiac output and hypotension. Due to the finding of pneumothorax on chest X-ray pigtail was introduced into the thoracic space. The diagnosis of total anomalous pulmonary vein drainage without visualization of confluence or vertical vein was suspected on echocardiography (ECHO). The patient underwent urgent open-heart surgery (derivation of pulmonary veins to left atrium using sutureless technique). Perioperatively, the presence of 2 right pulmonary veins and 1 left was

confirmed. Macroscopically, the lung had irregular surface with scattered nodular changes. After surgery, the patient was on ECMO due to severe pulmonary hypertension. A subsequent chest radiograph obtained after surgery showed persistent pneumothorax in the left and reduction of respiratory surface area in the right (Fig. 1). Realised invasive catheterisation showed relatively well developed lower pulmonary veins, but upper pulmonary veins were rudimentary. Pulmonary vasculature on the right side was poor (Fig. 2). The bronchoscopy excluded an obstruction of airways and smaller amount of mucus plugs was aspirated. High-resolution CT demonstrated diffuse interstitial changes involving thickening of the interlobular septa, which was associated with the presence of atelectasia (Fig. 3). Pulmonary hypertension persisted despite

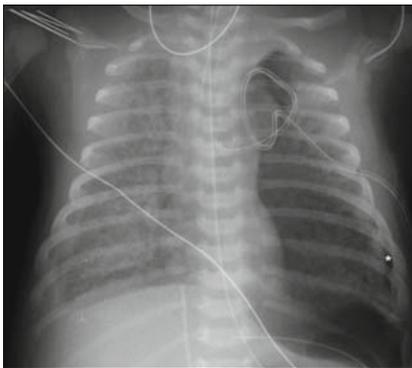


Fig. 1. Chest radiograph. Diffuse, bilateral nodular changes are present. There is a pneumothorax in the left (asterisk).

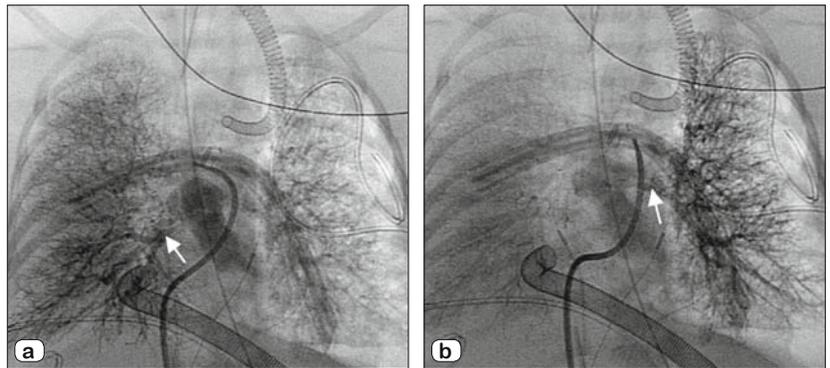


Fig. 2 a,b. Pulmoangiography with visualization of pulmonary veins. Arrows show relatively well developed lower pulmonary veins, but upper pulmonary veins are rudimentary (arrowheads).

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adequate therapy. His medical conditions started to worsen. He died of cardiac shock. Autopsy confirmed the diagnosis of congenital pulmonary lymphangiectasia (PL) (Fig. 4).

PL is a rare disorder involving the lung and is characterized by pulmonary subpleural, interlobar, perivascular and peribronchial lymphatic dilatation. PL presents at birth with severe respiratory distress, tachypnea and cyanosis, with a very high mortality rate

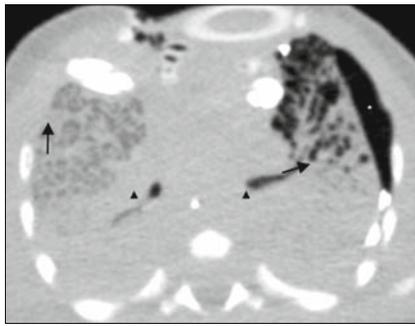


Fig. 3. High-resolution computed tomography (HRCT). There is a diffuse thickening of peribronchovascular interstitium and the interlobular septa (arrows), pneumothorax in the left (asterisk) and bilateral peribronchovascular infiltrates (arrowheads).

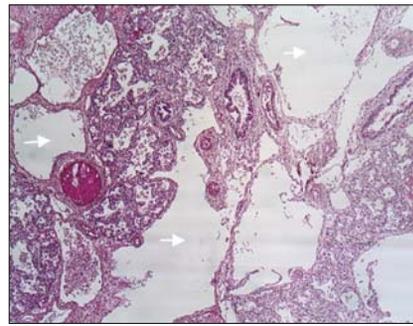


Fig. 4. Microscopical examination. Arrows show dilated lymphatic spaces.

at or within a few hours of birth. The etiology of this disease is not known. Most likely, PL lymphatic channels of the fetal lung do not undergo the normal regression process at 20 weeks of gestation. Secondary, PL may be caused by a cardiac lesion – pulmonary lymphatics dilatation develops in utero as a result of obstructed pulmonary venous flow. The most frequent causes of secondary PL are hypoplastic left heart syndrome, pulmonary vein atresia, congenital mitral stenosis, cor triatum, and thoracic duct agenesis. The diagnostic approach includes complete history, conventional radiographs, ultrasound and magnetic resonance examination, lymphoscintigraphy, lung functional tests, lung biopsy, bronchoscopy, and pleural effusion evaluation (especially if chylous). Fetal ultrasound examination plays an important role in the antenatal diagnosis of PL. Current advances in intensive neonatal care have

changed the previously fatal outcome of PL at birth. Patients with PL who survive infancy, present with medical problems which are characteristic of chronic lung disease (1, 2, 3).

References

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