An Autopsy Case of Alveolar Capillary Dysplasia
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Patient
Chief complaint: A full term, newborn girl developed respiratory distress shortly after birth

Hospital course: The term baby developed respiratory distress and worsening cardiopulmonary function secondary to meconium aspiration. She received maximum cardiopulmonary support including the oscillator, nitric oxide, and vasoactive agents. Echocardiogram showed normal anatomy of cardiovascular system, patent ductus arteriosus. Despite maximum support, she continues to show a worsening trend in her oxygenation. She was started on Extracorporeal membrane oxygenation (ECMO) on the second day, and was separated 1 week later. Two weeks after birth, the patient had increased shunting with stimulation and acidosis requiring increased FiO2 and ventilatory support. Respiratory surfactant was given and despite aggressive treatment and resuscitation, the patient expired after 20 days.

Lab data
CBC
- WBC 21.5 B/L
- 87% granulocytes
- 6% lymphocytes
- 7% monocytes
- Hg 15.7 g/dL

BLOOD CULTURES
Staphylococcus Aureus, Tracheal Aspirates
E. coli and Staphylococcus Aureus
Arterial Blood Gas
pH 7.4
Total CO2 19.0
PO2 81

Autopsy Findings

Discussions
This 20-day-old newborn baby was born at full term. Since birth, he had struggled with severe respiratory distress. Despite aggressive treatment including steroid, antibiotics, ECMO, and surfactant, he expired. Significant pathologic findings of the lungs are revealed as alveolar capillary dysplasia (ACD), with bronchopneumonia, focal infarction, and diffuse alveolar damage. The findings of patent ductus arteriosus and patent foramen ovale, are consistent with the patient’s clinical course of persistent fetal circulation, which is associated with persistent pulmonary hypertension caused by poor development of alveolar capillary and muscular thickening in the small pulmonary arteries. His pulmonary infections are associated with staphylococcus aureus which was detected in both blood and sputum.

ACD is an uncommon cause of irreversible persistent pulmonary hypertension in full-term newborn. In ACD, there is a failure of formation of air-blood barrier in addition to misalignment of pulmonary veins, which is also referred to as alveolar capillary dysplasia with misalignment of the pulmonary veins (ACD/MPV) (1). The incidence of ACD/MPV is not yet known, as the definitive diagnosis of ACD/MPV currently depends on histological examination of lung tissue. Gene changes in Forkhead Box F1 (FOXF1), a transcription factor gene critical for vascular development, are believed to be responsible for the pathogenesis of ACD based on gene sequencing in a cohort of 18 patients with the disease (2) and gene sequencing in a cohort of 18 patients with the disease (2) and animal models of ACD (3). Although typically fatal, a recent study (4) proposed that the severity of disruption in capillary density and the lack of contact with the alveolar epithelium may be two major factors that can determine clinical course and prognosis. Therefore recognition of this entity in a lung biopsy following clinical findings of persistent fetal circulation is important, because it may contribute to making the best medical decision.

References