A case of fetal chylothorax due to primary pulmonary lymphangiectasis: A postmortem diagnosis
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Objective
Primary pulmonary lymphangiectasis (PPL) is a rare congenital developmental abnormality of the lung with a generally poor prognosis. It includes hydrops fetalis, chylous ascites, intestinal lymphangiectasis, pleural and pericardial effusions, and pulmonary lymphangiectasis. The aim of this study is to report a case of PPL which was confirmed after postmortem analysis.

Methods
We report a case of antenatal chylothorax due to PPL diagnosed by ultrasound examination in Hospital das Clinicas of University of São Paulo.

Results
A 22-year-old woman of 27 weeks gestation, was referred to us due to antenatal ultrasound diagnosis of fetal asymmetric pleural effusion (mostly right), associated with ascites, hydrocele and mild subcutaneous edema. TORCH tests were performed in fetal and maternal samples, as well as karyotype analysis of amniotic fluid. No abnormalities were noticed in any of these exams. In order to relieve the mediastinal compression at 28 weeks of pregnancy, we proceeded to thoracoamniotic shunting under ultrasound guidance using a 14G trocar and a pigtail shunt. Migration of the shunt occurred one day after the procedure with rapid reaccumulation of pleural effusion. Due to the unfavourable fetal and placental (anterior) position, four thoracic drainages were performed with 18G needle in the subsequent week. Cytological analysis of the aspirate demonstrated a high percentage of lymphocytes (91% of the cells) suggesting the diagnosis of chylothorax. A cesarean section was performed at 30 weeks and 6 days, due to threatened preterm labor. The newborn (male, 2620g, Apgar 4/4) had a thoracocentesis and endotracheal intubation in the delivery room, but unfortunately succumbed to heart failure one hour after birth. Postmortem analysis revealed systemic dilation of the lymphatic vessels of the lungs, liver and kidneys, suggestive of PPL.

Conclusion
The true incidence of PPL is difficult to estimate as there has only been case reports and/or small case series in the literature. Further investigations should always be performed in cases of non-immune hydrops, because PPL is a rare, under-diagnosed complication with high mortality rate. Appropriate prenatal counselling, including documentation of maternal and fetal risks, and the decision to deliver should be managed by a multidisciplinary team.