Letters to the Editor

Fetal pulmonary thrombosis

We report a rare case of fetal pulmonary thrombosis with a structurally normal heart. A 44-year-old woman, gravida 2 para 1, was referred to our center at 23 weeks' gestation. Fetal ultrasonography revealed hydrops with bilateral pleural effusion, pericardial effusion, skin edema and ascites. The fetus had a structurally normal heart and no extracardiac anomalies. Thoracentesis was performed at 25 + 4 weeks' gestation. The cell content of the fetal pleural fluid revealed a lymphocyte percentage of 99%, suggesting chylothorax.

Thoracoamniotic shunting was performed using a double-basket catheter (Hakko Co., Japan) at 26 + 2 weeks' gestation. After a third shunting at 29 + 3 weeks, uterine contractions could not be suppressed by a combination of ritodrine hydrochloride and magnesium sulfate. Following adequate counseling, indomethacin as a suppository was used for tocolysis at a dose of 50 mg every 8 h. Three days later, the right atrium and ventricle of the fetus gradually dilated, and mild tricuspid regurgitation was observed. Indomethacin treatment was discontinued immediately, however, markedly decreased blood flow in the pulmonary artery and the ductus arteriosus were sequentially demonstrated by Doppler echocardiography at 30 + 1 weeks (Figure 1). A dilated main pulmonary artery with mild valve regurgitation was also observed. Although fetal hydrops markedly improved after a fourth shunt procedure at 30 + 4 weeks, blood flow in the pulmonary artery and the ductus arteriosus did not resume, and right ventricular failure did not improve. At 31 + 3 weeks the mother went into labor and an emergency Cesarean section was performed.

Figure 1 Color Doppler ultrasound image of a fetus with chylothorax and hydrops at 30 weeks' gestation, showing absence of blood flow in the pulmonary artery (PA) and ductus arteriosus. Dilatation of the main PA and mild pulmonary regurgitation (arrow, PR) can also be seen. LA, left atrium; RA, right atrium; RVOT, right ventricular outflow tract.
with delivery of a 1686 g male infant. The neonate died 3 h after birth as a result of hypoxia and metabolic acidosis.

Postmortem examination revealed a hypertrophic right ventricular wall and a severely dilated main pulmonary artery due to occlusion by an organizing thrombus (Figure 2a). The ductus arteriosus was not occluded, but was constricted. A mural thrombus within the dilated pulmonary artery had adhered to the arterial wall in a part that was already organized, as shown by Masson’s trichrome staining (Figure 2b). Excluding the stalk, the thrombus was not fully organized. This strongly suggested that the thrombus originated on the surface of the intima of the main pulmonary artery in the prenatal period. The pulmonary artery itself was structurally normal. However, focal CD34-positive endothelial cells were lacking.

We postulate that thrombus formation in this case was attributable to Virchow’s triad. In 1856, Rudolf Virchow proposed that abnormalities in blood flow, hypercoagulability of the blood, and injury to the vessel wall are causally related to thrombus formation. In our case, it is likely that reduced right heart ejection fraction caused by fetal hydrops and premature constriction of the ductus arteriosus after administration of indomethacin led to abnormalities in blood flow. Postmortem examination showed a focal defect of the endothelium of the fetal pulmonary artery. The multiple thoracoamniotic shunting procedures performed for fetal chylothorax might have caused hypercoagulability of the blood and injury to the vessel wall. We speculate that surgical stress caused inflammation leading to activation of leukocytes, thrombocytes and coagulation factors. We suggest that indomethacin should be used cautiously when repeated thoracoamniotic shunting is required for fetal chylothorax with hydrops.

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