Retroperitoneal lymphangioma

Holub M.[1], Smetanová D.[1], Hejtmánková M.[1], Prosová B.[2], Tomek V.[3]
1) Gennet, Center for Fetal Medicine and Reproductive Genetics, Prague, Czech Republic
2) Department of Radiology, Motol University Hospital, Prague, Czech Republic
3) Department of Cardiology, Motol University Hospital, Prague, Czech Republic

Abstract
We report the case of fetal lymphangioma located in the retroperitoneum. It was diagnosed by ultrasonography and magnetic resonance imaging (MRI) in 23 weeks of gestation.

Objective
Lymphangiomas are rare congenital malformations of the lymphatic system. The incidence of lymphangioma was reported to be 1:6000 at birth and 1:750 among spontaneous abortions. We had 21 cases in the Czech Republic from years 2009 to 2020. They arise due to a developmental defect in the lymphatic pathways and lead to accumulation of lymph. The lymphatic system of the embryo is supposed to develop around the fifth to the sixth week of gestation. Lymphangiomas can be divided into capillary, cavernous and cystic. The most frequent localization is the neck (75%), axilla (20%), and other organs (4%). They are associated with trisomies 13, 18, and 21, Turner syndrome, Noonan syndrome, hydrops and structural anomalies. The diagnosis is based on the ultrasound examination and MRI examination. Sonographically, we distinguish septated or nonseptated lymphangiomas. In examination algorithm we have to include fetal karyotype testing and consultation with the geneticists. Prognosis of fetuses with lymphangioma is unpredictable and depends on many factors. The treatment od lymphangioma includes surgical extirpation, medication and a careful follow up, however, expectant treatment is possible too. Lymphangioma causes fetal death in >50% of the cases. Incidence of relapse after treatment is 13%.

Results
We present a case of a 37 year old woman with a fetus in 23+3 weeks gestation. She was gravida 3, para 1. The medical history of the woman was normal. She was not taking any medication. She underwent an ultrasound scan in 23 weeks of gestation, which showed a multi cystic mass on the left side of the pelvis, which continued to the abdomen cavity. The size of the structure was 39x25x27mm. Left kidney was dislocated cranial by it. The umbilical vein was dilatated. AMC was performed with normal array results. The fetal ECHO was normal, without any obstructions in venous flow. The MRI confirmed our UZ examination. The main mass of the lymphangioma was on the left side of the retroperitoneum, spreading to the presacral area and even to the lumbal area and to the left thigh.

Conclusion
The aim of the case report is to present this rare disease. Like in other similar cases, our patient decided to terminate the pregnancy. The pathological examination confirmed all our examinations.