ÖZET
Yaygın fetal kistik lenfanjioma lenfatik sistem nadir bir konjenital malfarmasyonudur. Lenfanjiomanın prognozu; lezyonun boyutu, lokalizasyonu ve birlikte olduğu diğer anomalilerin varlığına bağlıdır. Olgumuzda maternal yaş 35, fetal BPD 27 hafta idi. Sunduğumuz bilateral proksimal ve distal alt ekstremite ve pelvik bölgeleri yaygın fetal kistik lenfanjioma olgusunda, pelvik bölgeden başlayan, her iki alt ekstremiteden distal kademede devam eden cilt altı yüzey ve derin yumuşak dokulara geniş kapsamlı olarak yaygınlık göstermiştir. Literatür araştırımlarımızda, her iki alt ekstremiteyi tutan sunulmuş bir lenfanjioma olgusu rastlanmamıştır.

Anahtar kelimeler: Fetüs; Lenfanjiyoma; Ultrasonografi

ABSTRACT
Extensive fetal cystic lymphangioma is a rare congenital malformation of the lymphatic system. Their prognosis depends on the size and location of the lesions as well as other accompanying anomalies. Our case was 35 years, fetal BPD was 27 weeks. Herein, we present a case of extensive fetal cystic lymphangioma that began at the pelvic area and symmetrically spanned the bilateral proximal and distal lower extremities. Numerous extensive and sharply circumscribed, thin walled multilobular cystic lesions in different sizes were observed in the subcutaneous superficial and deep soft tissue beginning from the pelvic area and extending to both lower extremities to the distal in the fetus. To our knowledge, a case involving both extremities has not yet been reported in the literature.

Key Words: Fetus; Lymphangioma; Ultrasonography

INTRODUCTION
Lymphangiomas are hamartomas of the lymphatic veins that can potentially infiltrate the surrounding structures. Approximately 50% of lymphangiomas are present at birth, and more than 90% become apparent by 2 years of age (1). Their prognosis depends on the size and location of the lesions as well as other accompanying anomalies. Lymphangiomas are frequently seen with polyhydramnios, hydrops fetalis and other anomalies, including skin edema and chromosomal anomalies. Prenatal diagnosis of these anomalies is of utmost. The neonatal consequences of extensive lymphangioma are generally malignant (2). However, spontaneous regression may occur in cases of fetal lymphangioma with normal chromosomes (3). The prenatal diagnosis of lymphangioma with ultrasound is important for planning the birth, timely postnatal resuscitation, treatment, and prognosis (3).

Herein, we present a case of extensive cystic lymphangioma that was identified in the 27th gestational week covers the pelvic area and the bilateral lower extremities.

CASE PRESENTATION
The maternal age was 35 years, the gravity was 6, the parity was 6, and the in vivo 6 BPD- FL (biparietal diameter-femur length) non-conformity was present in the fetus. The fetal BPD was 27 weeks/0 days, FL was 22 weeks/3 days and the AC (abdominal circumference) was 24 weeks/6 days 8 (682 gr +/-100 gr).

Numerous extensive and sharply circumscribed, thin walled multilobular cystic lesions in different sizes were observed in the subcutaneous superficial and deep soft tissue beginning from the pelvic area and extending to both lower extremities to the distal in the fetus (Figure 1, 2). Doppler US did not detect any flow in the cysts. Furthermore, cystic dilatations were observed in the fetal umbilical vein (Figure 3). Extensive edema was significant in the skin in both lower extremities. Wide vascular areas with endothelial row, including lymphocytic aggregates, were observed in cystic mass cytology, which the diagnosis of cystic lymphangioma (Figure 4).
DISCUSSION

Lymphangiomas are benign hamartomas of the lymphatic system that consist of multiple dilated veins. These generally originate from defects in the growth of the lymphatic channels, which generally develop in the 6th week of gestation. Histologically, there are 3 main types of lymphangiomas, as follows (1).

Type 1: Simple lymphangioma, consisting of lymphatic capillaries (2),
Type 2: Cavernous lymphangioma, made up of larger lymphatic vessels with wider fibrous adventitia (3), and
Type 3: Cystic lymphangioma or (cystic hygroma), which consist of multiple cysts in sizes ranging from a few millimeters to several centimeters.

All three types of lymphangiomas can be present at the same time in the same lesion. The cysts generally include serous or cystic liquid, but if complicated, they can have bloody or purulent content (4). The sizes of these lesions can vary from small liquid collections to large cysts. Despite being benign, these lesions can compress neighboring vital organs (5). In utero, their characteristic sonographic appearance is thin walled multiseptated cysts. The liquid can be anechoic via US (ultrasound) or different internal echoes, and liquid-liquid levels can depend on bleeding and fibrin deposition (6).

While 75% of lesions are located in the head-neck or the axilla, 25% are located in the trunk, 11% in the extremity, 11% in the mediastinum, 1% in the abdomen, and 3% in the genital area. Abdominal lymphangioma are the most frequently seen location, followed by the small intestine mesentery and the in retro peritoneal (7).

Pelvic and extremity lymphangiomas are rarely. To our knowledge, only 4 cases diagnosed with fetal abdominal lymphangioma partially extending to the extremities have been presented (2,4,7,8). We have not come across a lymphangioma case involving both extremities in the English literature.

In our lymphangioma case, involvement was present in both the pelvic area and in the proximal and distal sections of both lower extremities, which has not previously been presented in the literature. Our case underwent a medical abortion in the 30th week, and the diagnosis of cystic lymphangioma was confirmed with histopathology and cytology.

The pregnancy was terminated due to a bad prognosis based on three other cases in the literature (2,7).

Ultrafast MR (Magnetic resonance) screening in the prenatal diagnosis for this type of patients has become useful in the presentation of complex fetal anomalies (8). However, MR
screening was not conducted since our patient did not approve of the technique. US has been reported to be an important tool for the diagnosis of lymphangioma, for the determination of the size and growth rate of the tumor, and for the determination of the prognosis (2). Moreover, US is an easily accessible, non-invasive, accountable and inexpensive diagnosis method that can provide important information to the patient and his/her family regarding future examinations, planning, and prognosis.

REFERENCES