Cervical lymphangioma: a case report

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Abstract

Objective: We aimed to discuss the management of a cervical lymphangioma case which was prenatally diagnosed.

Case: A 22-year-old pregnant woman, gravida 1, parity 0, pregnant was referred to our clinic due to high risk detected during the second trimester screening tests. In the evaluation of fetus, a septate cystic mass that was measured as 37x34 cm and located in the left side of the neck was observed ultrasonographically. When performed Doppler ultrasonography to cystic mass, no blood flow was monitored. Other system examinations showed no additional anomalies. The cystic mass has grown up in the follow up of the fetus whose karyotype analysis was normal. The diagnosis of lymphangioma was confirmed pathologically after the postpartum excision of the cystic mass.

Conclusion: The presence of other anomalies should be investigated and karyotype analysis should be performed for the management of the cases of cervical lymphangioma. In isolated cases of cervical lymphangioma, the prognosis is good.

Key words: Lymphangioma, prenatal diagnosis, management.

Introduction

Cystic lymphangiomas are benign congenital lesions and they are the result of the non-existence of lymphatic canal system development or the existence of obstruction development. They are frequently located on neck, axilla, mediastinum, anterior abdominal wall and limbs. They may be with other structural and chromosomal anomalies as well as being isolated. Cystic lymphangiomas generally have a rapid growth pattern and they may perform local invasion and compression onto the tissue below. Also, intrauterine regression may be seen in 10-15% of cystic lymphangioma cases.1-3

It has been aimed to discuss the presentation and management of fetal cervical lymphangioma case which was prenatally diagnosed.

Case Report

A 22-year-old pregnant woman, gravida 1, parity 0, pregnant was referred to our clinic due to high risk detected during the second trimester screening test at 21 weeks of gestation. In the evaluation of fetus, a septate cystic mass that was measured as 37x34 cm and located in the left side of the neck was observed ultrasonographically (Figs. 1 and 2). No blood flow was observed dur-
ing the Doppler ultrasonography examination of the cystic mass (Fig. 3). No additional anomaly was detected during the examination of other systems.

The family was informed about screening test and examination findings of fetus, and karyotype analysis was recommended. In the follow-up of the fetus which had normal result for karyotype analysis, it was found that cystic mass grew. Besides, no pathology was found in venous Doppler findings, and no hydrops findings were found during gestational follow-ups. As a result of the evaluation of the pregnant who admitted to our clinic when her pains began and her waters broke at 39 weeks of gestation, it was anticipated that dystocia might develop, and the pregnant was delivered by cesarean.

A cystic mass that was measured as 37x34 cm and located in the left side of the neck of girl baby, whose 1st and 5th minutes APGAR score were 8 and 10, was observed. It was confirmed that the cystic mass of the baby was lymphangioma, and the baby was discharged on postpartum second day since there was no compression finding. It was detected during the follow-up of the baby that the cystic mass reduced (40x30 mm) at the second week (Fig. 4). The cystic mass was excised on 20th day by plastic surgery clinic and lymphangioma diagnosis was confirmed pathologically.
Discussion

The most frequent type of lymphangiomas is the cystic lymphangioma. The frequency of cystic lymphangiomas is 1/6000 and it is most frequently located on neck region (75-80%).[3] Cystic lymphangiomas are the lesions which have unicocular or multicocular view from small to giant dimensions, and have no flow in Doppler ultrasonography examination. In the differential diagnosis, cystic hygroma, hemangioma, teratoma, and fetal goiter should be considered first. Cystic hygroma is characterized by a cystic structure filled with bilateral septation fluid and separated by thick fibrous band corresponding nuchal ligament in the middle. Giant cystic lesions filling all amniotic cavity at second trimester is characterized with especially cranium, neck and septic skin edema filled with fluid in upper body. In the end, effusion and hydrops can be observed in body cavities. Hemangioma can be defined as cystic lesions including blood flow especially in Doppler ultrasonography. Neck teratomas are defined as heterogeneous lesions including calcifications where both solid and cystic structures are seen together in the ultrasonography. Fetal goiter is the lesions which are always in solid structure and with regular margins.[4]

In our case, multicocular cystic lymphangioma localized at neck area and displaying intrauterine growth was found and no additional anomaly was detected. It is known that non-immune hydrops fetalis develops in 43-75% of cystic lymphangioma cases. Approximately in 30% of cases with developing hydrops, it has been reported that karyotype is normal but additional anomalies (cardiac, renal, digestive and skeletal system anomalies) accompany and prognosis of cases with developing cases is bad.[3,5] Also, it has been reported that it may cause cardiac failure by distorting venous flow through increasing lymph edema. In the follow-up of our case, no pathology has been detected in venous Doppler finding. It is reported in the literature that lymphangioma cases may be associated with trisomy 13, 18 and 21, Turner syndrome, Noonan syndrome and structural anomalies. It has been reported that Turner syndrome has been seen in 42% of the cases, trisomies in 18% of them, and normal karyotype in 38% of them.[6]

The karyotype was normal in the case that we presented. Prognosis is good in isolated cystic lymphangioma cases, and successful outcomes are achieved by surgical excision and sclerotherapy at neonatal period. It is also reported that intrauterine sclerotherapy is safe.[7] Also, in lymphangioma cases localized in face and neck regions, urgent tracheostomy or EXIT (extrauterine intrapartum treatment) procedures may be required due to the obstruction syndrome on congenital upper respiratory tract.[8,9]

Conclusion

In cervical lymphangioma cases, other structural anomalies should be researched and karyotype analysis should be recommended. Prognosis is good in isolated cases, but they should be followed up for fetus cardiac failure and hydrops.

Conflicts of Interest: No conflicts declared.

References