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Case Report

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The giant fetal axillary lymphangioma showing good prognosis

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Abstract

Objective: Lymphangiomas are the rare congenital malformations of lymphatic system. It generally develops on head, neck and axillary regions. It has three types which are capillary lymphangioma, cavernous lymphangioma and cystic lymphangioma. In this case, we aimed to present the case of giant cystic lymphangioma (cystic hygroma) which begins from right axillary region and involves thorax and abdomen who was diagnosed prenatally and had surgical treatment on neonatal period.

Case: 34-year-old pregnant woman who had to labor was admitted to our clinic. The ultrasound examination revealed a large multiseptated cystic mass measuring 129x92 mm in size in right axillary region. No fetal abnormalities were found by ultrasonography. Newborn had a successful surgery in the postpartum period.

Conclusion: Even if they are large axillary lymphangioma, prenatal follow up and postnatal treatment should be consider.

Key words: Fetal, cystic lymphangioma, prognosis.

İyi prognoz gösteren fetal dev aksiller lenfanjioma

Amaç: Lenfanjiomalar, lenfatik sistemin nadir konjenital malformasyonlardır. Genellikle baş, boyun ve aksiller bölgede gelişir. Kapiller lenfanjioma, kavernöz lenfanjioma ve kistik lenfanjioma olmak üzere üç tipi vardır. Bu olguda, prenatal olarak tanısı konulan ve neonatal dönemde cerrahi olarak tedavisi yapılan, sağ aksiller bölgeden başlayıp toraks ve abdomeni içine alan dev kistik lenfanjioma (kistik higroma) olgusunu sunmayı amaçladık.

Olgu: Doğum ağrıları olan 34 yaşında gebe kadın kliniğimize başvurdu. Ultrasonografik inceleme ile sağ aksiller bölgede, 129x92 mm boyutunda ölçülen büyük multiseptasyonlu kitle saptandı. Yapılan ultrasonografide herhangi bir fetal anomali izlenmedi. Yenidoğan postpartum dönemde başarılı cerrahi operasyon geçirdi.

Sonuç: Aksiller lenfanjiomlar büyük olsalar bile prenatal takip ve postnatal tedavi akılda tutulmalıdır.

Anahtar sözcükler: Fetal, kistik lenfanjiom, prognoz.

Introduction

Cystic lymphangioma (CL) develops depending on the connection between venous system and lymphatic vessels within soft connective tissue such as especially head-neck and axilla. It is observed in 1/200 of spontaneous abortions.[1-3] They can be accompanied by chromosomal anomalies frequently as well as being observed in an isolated condition. Intrauterine death is observed in 33% of these cases. Mortality is 100% in cases with hydrops and oligohydramnios. It is reported that 10-15% of intrauterine follow-ups have spontaneous regression.[4] The preferred treatment method is the total excision of cystic mass.[5,6] Final diagnosis is established after histopathological examination of the cystic mass. In our report, we aimed to present giant cystic lymphangioma case which reached term pregnancy, recovered by postnatal total excision and of whom diagnosis was proven pathologically.

Case Report

Thirty-four-year-old pregnant (gravida 3 and parity 2) who did not have routine pregnancy follow-up applied

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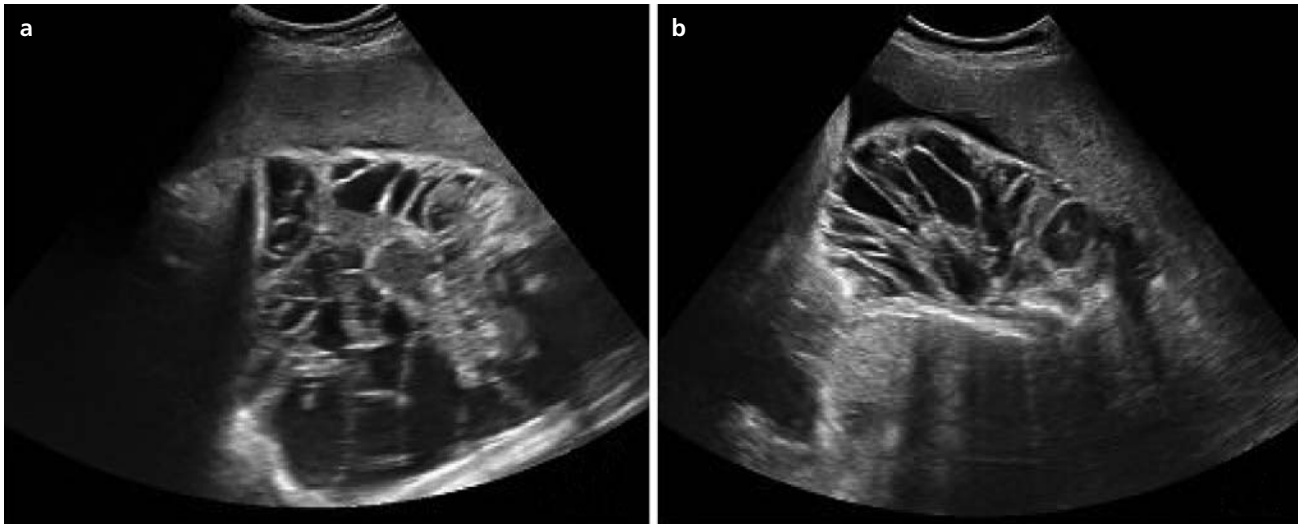

Figure 1. Ultrasonographic view of cystic lymphangioma of fetus on (**a**) sagittal and (**b**) coronary sections.

to our gynecology clinic when her labor pains began. There was no significant indication at her anamnesis. A single 38-week-fetus on head presentation with normal levels of amniotic fluid was observed in the obstetric ultrasonography. Placenta had a normal view. There was no apparent anomaly in the fetus except septated cystic mass measuring 129x92 mm in size beginning from right axillary region and involving thorax and abdomen (**Figure 1**). The patient who had active contractions was delivered by cesarean due to large baby indication with the concerns that cystic mass could be ruptured and dystocia could develop. The baby was female which was 4,370 gram in weight, 50 cm in length and having 1st and 5th minute Apgar scores as 4 and 8, respectively.

Consultation was asked on postnatal period from the departments of pediatrics and pediatric surgery. Genetic analysis was performed and the outcome was

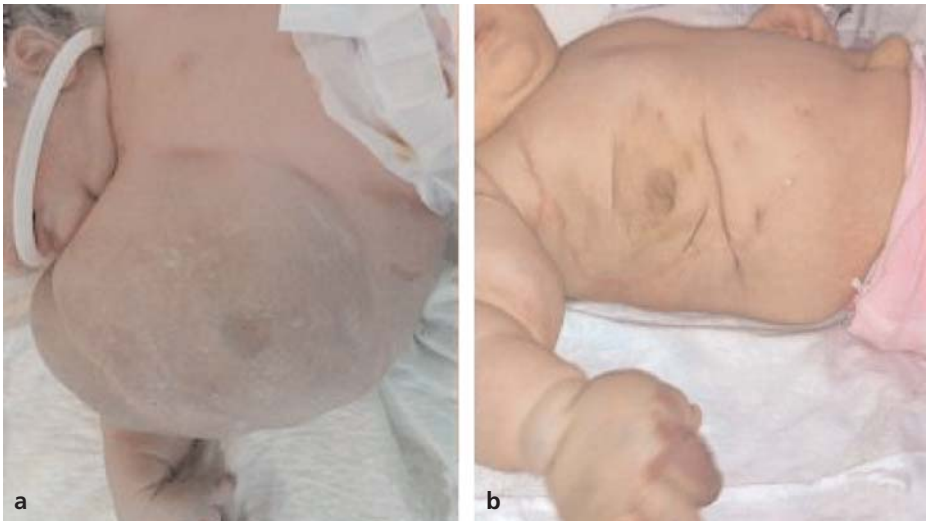

Figure 2. (**a**) Preoperative and (**b**) postoperative views of newborn with cystic lymphangioma.

reported as normal. A mass beginning from right axillary region and reaching towards anterior thoracic wall and downward was observed in the examination (**Figure 2a**). After ultrasonography and computed tomography examinations, a septated cystic mass was observed which was measuring about 88x105 mm in axial size beginning from right axillary region and reaching up to iliac crest, and where irregular limited heterogeneous and hyperdense areas existed under skin. No pathology was found in echocardiography. Near total mass excision was done to the baby followed-up in newborn intensive care unit when it was only 6-day-old by pediatric surgery department. Pathology result of the material was reported as lymphangioma (cystic lymphangioma) and one reactional lymph node. The baby followed up in newborn intensive care unit during postoperative period was discharged from the clinic with full recovery (**Figure 2b**). No pathology was observed in the baby who was controlled on postpartum 3rd month.

Discussion

Cystic lymphangiomas are seen in 1/6,000 of pregnancies and it is the most frequent type of lymphangiomas. There are localized and common forms. It may be seen mostly on neck region (75%) and especially on posterior region, and they may also reach to thorax, axillary region, abdomen, scrotum and bones.[4,7] In some cases, spontaneous regression is reported.[8] It is asserted that this regression is caused by increased pressure on lymphatic system which is able to pass the incomplete obstruction in especially non-septated cases.[9] In our case, a septated cystic mass was observed which was diagnosed as lymphangioma measuring 129x92 mm in size beginning from right axillary region and involving thorax and abdomen.

Cystic lymphangiomas may be isolated as well as accompanying a group of diseases where especially there are chromosomal anomalies. Turner syndrome is observed 40-80% of them. Turner syndrome is observed frequently in female fetuses with cystic lymphangiomas. They may sometimes present familial inheritance. Prenatal diagnosis tests for genetic scanning purposes were not done on the presented case since she did not refer to our clinic before. It was learnt from her history that she had no such anomaly in her other 2 children and her close relatives. No abnormal indication was found except giant cystic mass presenting elongation from right axillary to downward in the female baby who was delivered by cesarean due to large baby indication. Genetic analysis was performed on postnatal period.

The size and being septated of cystic lymphangiomas are bad prognostic factors and they generally accompany chromosomal anomaly.[9] According to some authors, septation has no prognostic significance.[10,11] In our case, there was both giant and septated wide CL according to ultrasonographic indications at admission time. However, we do not know the development phases of the mass since there is no ultrasonographic examination on early periods of the pregnancy.

At a rate of 43-75%, cystic lymphangioma may cause non-immune hydrops. It is asserted that high protein concentration within cyst causes common edema by creating hypoproteinemia.[10] Also increasing lymphedema may cause pleural, pericardial and abdominal effusion, and cardiac failure by distorting venous return. It is reported that oligohydramnios is developed in approximately 2/3 of cases. It is considered that the reason of oligohydramnios is fetal hypovolemia, hypoperfusion and secondarily decreased renal perfusion.[10] In our case, hydrops indications, abnormal placental view and oligohydramnios were not observed.

Postnatal echocardiography indications were evaluated as normal. It has become easy to diagnose on early periods of pregnancy by the use of high-resolution ultrasonography device. The prognosis of disease depends on gestational age, karyotype, presence of septations and accompanying anomalies during diagnosis.[12] In the presented case, gestational age reached the term, and the mass was big and septated. There was no accompanying anomaly. Since the patient did not refer at early pregnancy period, karyotyping could be done at postnatal period and it was reported as normal.

Conclusion

Lymphangiomas and accompanying anomalies can be recognized by ultrasonography. We consider that in pregnancies where there is no additional other anomaly, and abnormal karyotype analysis at prenatal genetic scan, hydrops indications and oligohydramnios, it should be taken into consideration the option of follow-up for especially axillary located lymphangiomas even they are giant and septated; and the option of terminating pregnancy after informing family in case of the presence of indications mentioned before viability limit.

Conflicts of Interest: No conflicts declared.

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