Prenatal Diagnosis of Fetal Chest Lymphangioma

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Objective. Prenatally diagnosed cystic nuchal hygroma is often associated with chromosomal anomalies and hydrops fetalis. Chest lymphangioma diagnosed later in gestation appears to be a completely different disease, with a low incidence of chromosomal and structural anomalies. Methods. Two chest cavernous lymphangiomas of the fetus are presented. The sonographic images, chromosomal analyses, and macroscopic and microscopic evaluations are described. Results. Fetal chest cavernous lymphangiomas were identified at 15 and 22 weeks' gestation. In the first case, the couple decided to interrupt gestation. In the second case, prenatal sonography showed a multilocular, cystic lymphangioma external to the chest wall with no flow on Doppler sonography. Follow-up sonography revealed normal fetal growth and slow enlargement of the cystic mass surrounding the left chest cavity. The neonate was delivered without complications and was treated surgically. Conclusions. The chest lymphangioma appears to be a lesion usually not associated with other congenital abnormalities. The prenatal diagnosis of chest wall lymphangioma is relatively easy sonographically, and the treatment of choice is surgical excision. The outcome is relatively favorable, with a low incidence of chromosomal and structural anomalies. Key words: chest lymphangioma; cystic hygroma; fetus; lymphangioma; lymphatic system.
Case Descriptions

Case 1
A 25-year-old woman, gravida 0, para 0, was referred at 15 weeks’ gestation for evaluation of fetal growth and an anatomic survey. Previous sonography at 12 weeks’ gestation was consistent with gestational age, and nuchal translucency measured 1 mm. A targeted sonographic examination revealed a single fetus with movements and amniotic fluid showing no abnormalities. The fetal head, spine, heart, abdomen, and extremities appeared normal. Two symmetric masses on the lateral aspects of the fetal chest could be identified. The masses were honeycombed in appearance, with multiple echo-free areas of varying size in the mass (Figure 1). The couple decided to interrupt the gestation because of the abnormal fetal finding. Chromosomal analysis was performed on amniotic cells, and the karyotype was found to be normal (46,XX). The sonographic findings were confirmed on postabortion examination (Figure 2). Macroscopic and microscopic evaluations revealed a fetal chest cavernous lymphangioma.

Figure 1. Axial image of the fetal chest at 15 weeks’ gestation, at the level of the 4-chamber view, showing a symmetric mass, honeycombed in appearance, with multiple echo-free areas of varying size in the mass.

Case 2
A 30-year-old woman, gravida 2, para 1, was referred at 22 weeks’ gestation for targeted evaluation of the fetal anatomy. Previous transvaginal sonography at 15 weeks’ gestation was consistent with gestational age, and the results of the anatomic survey were considered normal. Nuchal translucency measurement at 12 weeks’ gestation also appeared to be normal. Chromosomal analysis was performed, and the karyotype was found to be normal (46,XX). The 22-week scan demonstrated fetal organ measurements appropriate for gestational age, with the amount of amniotic fluid and the movements showing no abnormalities. The fetal head, spine, heart, abdomen, and extremities appeared normal. In addition, an abnormal mass connected to the fetal chest wall could be identified (Figures 3 and 4). The mass was unilateral and honeycombed in appearance, and echo-free areas were scattered inside the mass with no flow on Doppler sonography. The couple decided to continue the pregnancy. Chromosomal analysis was performed, and the karyotype was found to be normal (46,XX). Repeated sonographic examinations during gestation showed slow growth of the mass without invasion into the deep tissues. There was no involvement of the left hand or the axillary region. A fetal magnetic resonance imaging study at 29 weeks was consistent with the sonographic images. The patient was advised to undergo elective cesarean delivery, but she refused. She had an uneventful spontaneous vaginal delivery at 40 weeks’ gestation. It is assumed that the location of the tumor without shoulder involvement and its soft texture made the delivery of the fetal body uncomplicated. The sonographic findings were confirmed after birth (Figure 5). The mass was excised at the age of 1 month, and microscopic evaluation revealed a chest wall cavernous lymphangioma.

Discussion

Cystic hygromas are congenital lesions that have been described extensively in the obstetric literature. Approximately 75% to 80% of all cystic hygromas involve the neck. A nuchal cystic hygroma that appears early in gestation is characterized by thin-walled cysts in the posterior nuchal region of the fetus, accompanied by gen-
eralized hydrops and structural abnormalities in many cases. It is also associated with a high incidence of chromosomal abnormalities and an extremely poor fetal outcome.1–3 Chest wall lymphangioma, however, seems to be a completely different disease, and prenatal diagnosis of this condition is rare. The findings may be unilocular or multilocular, and the lesions range in size from several millimeters to much larger and contain a clear or cloudy lymphoid fluid. Lymphangiomas are believed to be caused by the anomalous development of the lymphatic system; the etiology is variable, probably multigenic. Lymphangiomas are made up of lymphatic vessels supported by connective tissue. No communication exists between the normal lymphatic system and the lymphangioma. Lymphangiomas have a predilection for local infiltration of the dermis, subcutaneous tissue, and soft tissue and occasionally are widespread.4,6

In contrast to cystic hygroma, chest lymphangioma may be a different congenital anomaly.6–8 Other authors5 have reported 11 cases of late-onset congenital cystic hygroma. It has been suggested that late-onset cystic hygroma should be differentiated from early-onset nuchal cystic hygroma because of the low incidence of chromosomal aberrations. McCoy et al9 reported a massive axillary cystic hygroma and normal karyotype, and they noted the possible difference in the etiology of prenatally nuchal cystic hygromas and cystic lymphangiomas in other anatomic locations. Our 2 cases had normal karyotypes.

Late-onset cystic hygroma is located mainly in the axilla and anterior abdominal wall.5–8 The short-term risk of this lesion is dystocia during labor, and the long-term risk may be the presence of an infiltrative microcystic component and organ involvement. In such cases, consequences are chronic lymphorrhea, infections, hemorrhages, and relapses of the lymphatic lesion leading to recurrent, difficult, and incomplete surgical procedures with the subsequent risk of functional impairment.9

The decision to terminate the pregnancy in our first case was very difficult to make. The normal karyotype and the absence of other anomalies argued in favor of maintaining the pregnancy; however, after detailed explanations, the couple decided to interrupt the pregnancy. In the second case, the decision to continue the pregnancy was made after a detailed explanation concerning the findings. Sonographic examinations showed slow growth of the mass but without invasion into the deep tissues. In addition, no other structures were involved, and the fetal magnetic resonance imaging study was consistent with the sonographic images.
The pathophysiologic features of lymphangiomas occurring in late gestation are unlikely to originate from lymphatic obstruction because the formation of lymph sacs is completed by the 6th to 9th weeks of pregnancy.1 The absence of associated structural and chromosomal abnormalities further suggests a different pathophysiologic mechanism. The clinical and obstetric consequences of these types of lymphatic lesions are very different.

We suggest that chest wall lymphangioma should be included in the entity of findings of low incidence for chromosomal abnormalities. These lesions are usually not associated with other congenital abnormalities or generalized lymphedema. The prenatal finding of chest wall lymphangioma is relatively simple and easy to diagnose sonographically, and the treatment of choice is surgical excision.10 The outcome is relatively favorable, with a recurrence rate of 10% to 15%, depending on the technical possibilities of complete removal of the pathologic tissue.

In conclusion, classic cystic hygroma is characterized by a high association with structural and chromosomal abnormalities, and the prognosis is extremely poor; however, chest lymphangioma appears to be a completely different disease, with a low incidence of chromosomal and structural anomalies, similar to late-onset cystic hygroma. Its outcome is relatively favorable. Other studies involving a greater number of patients are necessary to better understand the evolution and prognostic factors of fetal chest lymphangioma.

References
