Fetal cystic lymphangioma of the neck: a case report

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Introduction

Lymphangiomas are congenital malformations of the lymphatic system with thin-walled vessels. The incidence of lymphangioma was reported to be 1:6,000 at birth and 1:750 among spontaneous abortions (1).
The lymphoangioma is believed to be derived from abnormal transformation of lymphatic fluid of the jugular lymph sacs in various sonographic appearances.
Lymphoangioma are large swellings mainly in the inferolateral part of the neck and consist of large single or multilocular flui cavities. They are frequently associated with karyotypic abnormalities, various malformation syndromes, and several teratogenic agent. Cystic hygroma diagnosed after birth is usually associated with a good prognosis.
Sonographically, the lesion may be either septated or not (nonseptated) by internal trabeculae. Fetuses with septated lymphangioma were more likely to be aneuploid and shall have a poorer pregnancy outcome than fetuses with nonseptated lesions.
It is still a dilemma which risk factors are associated with perinatal outcome.

Case report

A 34-year-old primigravida presented with a fetus at 21+5 weeks' gestation with a neckl cystic mass. Her family history was negative. Ultrasound examination revealed a multicystic and hypoechogenic mass extending from antero-lateral neck region of 44 mm diameter (Fig. 1).
Color Doppler immage showed a modest vascolarizzation regular departure from the vascular bundle of the neck by which has also contiguity relationships in it (Fig. 2).

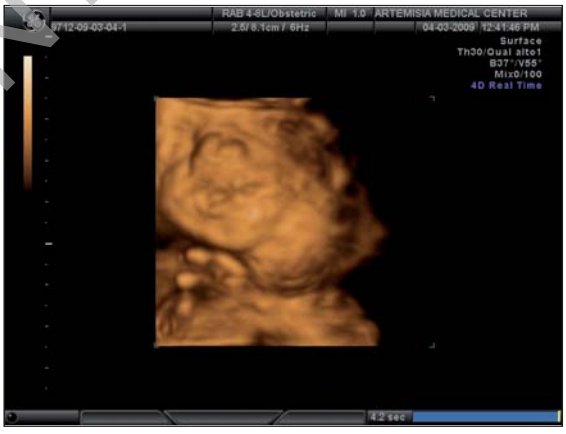

Figure 1

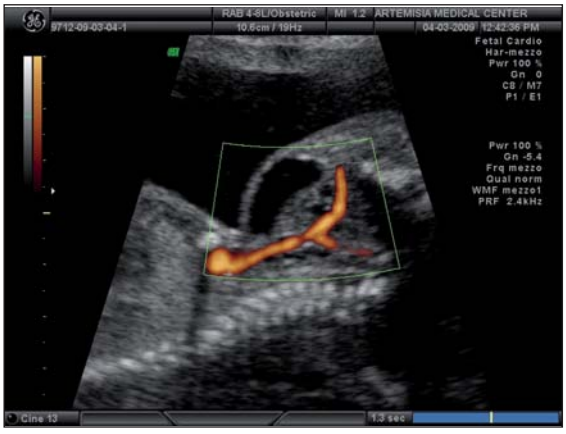

Figure 2

The trachea appeared not diverted, was not present polidramnios. The typical sonographic findings suggested a lymphangioma. Fetal biometry was appropriate for gestational age.
At 22 weeks' gestation, amniocentesis was performed and a chromosomal analysis revealed a normal 46, XY karyotype but was heterozygous for congenital deafness.
We explain to the patient that in most cases, the cystic lynphangioma is solved when karyotipe is negative. Infact the post natal success rate with treatment sclerosing can get, in the forms with single large cyst, 75%-80%. The incidence of relapse after surgery is estimated at 13%.
Our patient decided to terminate her pregnancy elsewhere, and we don't have feed back of the autoptical examination of the fetus.

Discussion

Lymphatic malformation is a benign, congenital proliferation of lymphatic tissue. The clinical term "cystic Hygromia" refers to a large bulky lesion containing cystic spaces, but this is not a histologic distinction (3).
Lymphangiomas are malformations of the lymphatic vessels is more frequent in the neck in the cervical region (75%) and constitute about 25% of all cysts of the neck. They may localize to both the front and rear triangle, and may be bilateral or monolotaral (4).
The lymphatic system of the embryo is supposed to develop around the fifth to sixth week of gestation. Six lymphatic sacs; two jugular sacs draining the head, neck, and arms; two iliac sacs draining the legs and lower trunk; and two sacs draining the gut called the cisterna chili, and the retroperitoneal sac, develop near the large veins .The lymphatic vessels develop from these sacs along the major veins. Two large channels (right and left thoracic duct) connect with the venous system at the junction of the internal jugular and subclavian veins near the end of the sixth week of gestation . Failure of the lymphatic system to connect and drain into the jugular veins leads to lymphatic fluid stasis, dilated lymphatic vessels and varying fluid-filled masses called the lymphatic obstruction sequence. If the connection between the jugular lymphatic and venous system, or an alternative route is established, lymphangiomas might theoretically resolve. We can have four type of lymphangioma:
I type: tumors which had no or only a minimal effect on the contour of the neck
II type: lymphangiomas were smaller than a line drawn at the lateral border of the head
III type: tumors exceeded this line
IV type: the lymphangiomas extended beyond the midline of the body.
During prenatal life the incidence and diagnosis of linfangiomi is not yet established, given the difficulty of diagnosis (7).
This fetal malformations can also produce neonatal airway obstruction. Prenatal sonographic diagnosis of these lesionsm permits anticipation of airway obstruction before birth and enables the development of a treatment strategy for effective perinatal airway management.
Lymphangiomas are known to be associated with trisomies 13, 18, and 21, Turner syndrome, Noonan syndrome, hydrops, structural anomalies and intrauterine demize. In our case, the karyotype was normal but the pregnant decided to terminate her pregnancy at 25 weeks' gestation on the base of heterozygosity of congenital deafness despite several apparently favorable prognostic features (atypical location, normal karyotype. The review of the letterature for cystic hygromas show that 42% of infants are 45,X, 38% have a normal karyotype, and 18% have trisomies. Prognosis remains guarded regardless of all other factors until the fetus reaches 26 weeks' gestation, after which time a 67% chance of ultimate survival can be expected. Only 42% of documented survivors were completely normal at follow-up.
Ultrasound, computed tomography, and magnetic resonance imaging are useful in confirming the diagnosis of lymphatic malformation (9). They are also helpful in delineating the extent of the mass and revealing its relationship to the surrounding structures. Furthermore, imaging will suggest the presence of any hemangiomatous component or large blood vessels. This will allow the surgeon to plan an operative approach to these lesions.

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