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PRENATAL ULTRASOUND DIAGNOSIS AND FOLLOW UP OF A RARE CASE OF A LYMPHANGIOMA LOCATED IN THE FETAL THORAX AND NECK WITH PROGRESSIVE ENLARGEMENT

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ABSTRACT:

Fetal lymphangiomas or cystic hygromas are uncommon congenital malformations of the lymphatic system. Prenatal diagnosis of lymphangiomas allows appropriate management planning, regarding pregnancy, delivery and treatment. We present a rare case of extended fetal lymphangioma, diagnosed on routine second trimester anomaly scan, initially located in the right mediastinum, with extension to the right lateral part of the neck. Molecular analysis using aCGH after amniocentesis showed a normal female karyotype 46,XX. The lymphangioma gradually enlarged and by the end of the third trimester it extended to the anterior and posterior part of the neck, left mediastinum, pericardium and right axilla. Elective cesarean section in a tertiary center allowed immediate evaluation by a specialized team of pediatric surgeons. Subsequent fetal MRI confirmed prenatal ultrasound findings. Thus far, based on the location and size of the lesion and the lack of pressure effects on the heart and the vital anatomical structures of the mediastinum, the lymphangioma has been treated with repeated courses of sclerotherapy only, which led to partial regression. Today, the child is a thriving 4.5 year old girl with normal physical and mental development.

Introduction

Fetal lymphangiomas or cystic hygromas are uncommon congenital malformations of the lymphatic system. They are characteristic thin-walled cystic dilations, most commonly found in the posterior neck [1]. Occasionally, they may be located in the mediastinum; most of the mediastinal cystic hygromas are extensions of cervical lesions, while lymphangiomas confined solely to the mediastinum are very rarely encountered [2]. Extension from the neck to the axillary region may also occur [3]. Enlargement of these cystic lesions is not uncommon, leading to possible compression of adjacent organs, including postnatal respiratory distress, feeding difficulties and/or vascular compromise. We present a rare case of a large fetal lymphangioma, diagnosed on routine second trimester anomaly scan in the right mediastinum and fetal neck, with progressive enlargement.

Case Report

A 29 year old white woman (G1, P0) presented for routine second trimester anomaly scan. Measurement of nuchal translucency in the first trimester was normal and the first trimester combined test was low risk for Down syndrome. Ultrasound revealed a cystic lesion in the right mediastinum, with few internal septums. Its diameter was 17 mm and the lesion was surrounding the heart and large vessels. Given the absence of blood flow on color Doppler, lymphangioma was the most likely diagnosis. The lesion extended to the right lateral part of the neck reaching a longitudinal diameter of approximately 4 cm (Figure 1A-C). After detailed counseling and signed informed consent, amniocentesis was performed and aCGH analysis revealed a female fetus with a normal 46,XX karyotype. Molecular genetic analysis of chromosomal regions and 120 detectable by aCGH genetic syndromes, showed no association with any detectable abnormality. Fetal echocardiography at 23 weeks was normal. Fetal MRI was not performed due to maternal contraindications (the mother had a cochlear implant). Follow up ultrasound scans were performed regularly until 37 weeks; there were no signs of fetal hydrops; however the mass was expanding in the following order: anterior and posterior part of the neck of the fetus, left mediastinum, pericardium and right axilla (Figure 1D-F). A multidisciplinary team extensively counseled the patient, including advice from an obstetrician and a pediatric surgeon regarding the mode of delivery and further management. An uneventful cesarean section was performed at 39 weeks in a tertiary center. Postnatally, the infant was stable and MRI findings were consistent with those of prenatal ultrasound. Repeated courses of intralesional injections of sclerosing agents (bleomycin) were administered, leading to successful reduction in size of the lymphangioma. Interestingly, the partial regression of the lymphangioma occurred in the reverse order of its intrauterine expansion, in the following order: right axilla, pericardium, left mediastinum and anterior part of the neck. The lymphangioma still persists in its primary location (right mediastinum and lateral part of the neck). Today, the child is a thriving 4.5 year old girl with normal physical and mental development.

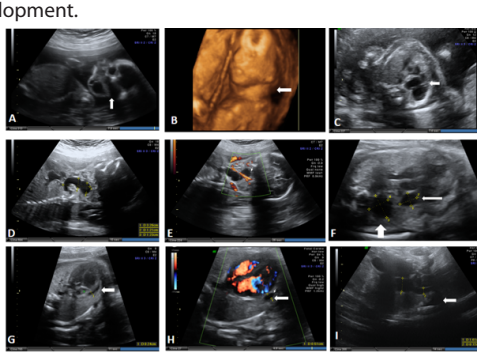

Figure 1. A and B: Two- and three-dimensional views of right lateral lymphangioma of the neck, at 21 weeks' gestation (arrows). C: Transverse view of the lymphangioma in the right mediastinum (arrow) at the level of the three vessel view, at 21 weeks. D: At 27 weeks, the septated lymphangioma extended from the right mediastinum to the right lateral and anterior part of the neck. E: On color Doppler, there was no blood flow within the lymphangioma, but there was infiltration of the adjacent vessels of the neck (27 weeks' gestation). F: Three-vessel view showing the lymphangioma (thick arrow) and dilatation of the right superior vena cava (thin arrow) at 27 weeks. G and H: Expansion of the lymphangioma to the pericardium (arrows) at 33 weeks. I: Expansion of the lymphangioma to the right axilla (arrows) at 33 weeks.

Discussion

Fetal lymphangiomas or cystic hygromas are uncommon congenital malformations, typically detected between the late first and early second trimester. Their incidence is 1 in 6,000 pregnancies, with most occurring in the neck and axilla; lymphangiomas confined to the mediastinum are quite rare and most remain asymptomatic until adulthood [3, 4]. Lymphangiomas are commonly associated with chromosomal abnormalities and poor outcomes; 9% occur in healthy children, while 2% occur in liveborns but with chromosomal anomalies or various malformations [5].

The postnatal outcome depends on the size and location of the lesion. Isolated lymphangiomas generally have a favorable prognosis and sclerotherapy or surgical resection is effective in most cases. On ultrasound and MRI, lymphangiomas are unilocular or multilocular cystic masses, with thin or thick walled septa. On ultrasound, the fluid may be anechoic or with variable internal echoes, due to bleeding and fibrin deposition. The absence of blood flow on color Doppler is characteristic of lymphangiomas in contrast to hemangiomas [6]. In the present case, second trimester ultrasound and absence of blood flow on color Doppler allowed accurate prenatal diagnosis. Furthermore, serial prenatal ultrasound examinations allowed accurate monitoring of the progressive enlargement of the lesion through the second and third trimester and planning of delivery and postnatal management.

Due to the complications of surgical treatment and high incidence of morbidity and recurrence, alternative treatments such as sclerotherapy, laser therapy and IFN-alpha systemic therapy have also been used in selected patients. Recent advances in sclerotherapy have expanded the contemporary lymphangioma management options. Sclerotherapy is considered appropriate for lesions that are not surgically resectable and it is also useful in reducing the size of a lymphangioma prior to surgical removal. Intralesional bleomycin, sclerotherapy with OK-432 (streptococcal derivate) or percutaneous embolisation with Ethibloc have all been reported to be effective [7, 8]. Although spontaneous regression of fetal lymphangiomas may occur, extensive lymphangiomas require a team approach in the post natal period. Recurrences are known to occur in one third of the cases, regardless of the size of lesion and type of the treatment [8, 9].

Prenatal ultrasound examination plays an important role in prenatal diagnosis and counseling regarding prognosis and treatment, as well as in guiding intrauterine operative treatment. Indeed, promising results with intrauterine treatment of a cystic hygroma with OK-432, a lyophilized mixture of Group A Streptococcus pyogenes and benzyl penicillin 432 have been reported [10, 11]. However, Ogita et al. [12] reported two cases of failure in large septated tumors.

In conclusion, a rare case of a large fetal lymphangioma diagnosed on routine second trimester anomaly scan initially located in the right mediastinum and the right lateral part of the neck which gradually became larger, expanding to the anterior and posterior part of the neck of the fetus, left mediastinum, pericardium and right axilla is reported. Serial prenatal ultrasound examinations allowed accurate monitoring of the gradual enlargement of the lesion appropriate planning of delivery and management immediately after birth. Though surgery is usually the most appropriate treatment, conservative management with sclerotherapy is an acceptable option in stable patients without unfavorable pressure effects on the heart and the vital anatomical structures of the mediastinum. Interestingly, the regression of the lymphangioma in the present case occurred in the reverse order of its intrauterine expansion.

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