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Case Report

## Fetal axillary lymphangioma

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### ABSTRACT

Lymphangioma is a rare congenital malformation of the lymphatic system that occur in 1 per 6000 live births. The most common site of lymphangioma is the neck (also named cystic hygroma), accounting for 75% of the cases, and it is strongly related to aneuploidies. Axillary location is very rare and appears not to be associated with chromosomal abnormalities. Tumor growth, fetal anemia secondary to intralesional bleeding, hydrops fetalis and shoulder dystocia are possible obstetric complications of this condition. The prognosis is generally good in the absence of abnormal karyotype, fetal hydrops or extension of the lymphangioma to adjacent tissues. Surgical excision or sclerotherapy are the main treatment choices. The authors present a case of a right fetal axillary lymphangioma and review the literature.

**Keywords:** Axillary lymphangioma, Prenatal diagnosis, Ultrasound

### INTRODUCTION

Lymphangioma is a rare congenital malformation of the lymphatic system that occur in 1 per 6000 live births.<sup>1,2</sup> Around 75% of the lymphangiomas occur in the neck and are usually named as cystic hygromas.<sup>3</sup> Other locations include the axilla (20%), the retroperitoneum and the intra-abdominal organs (2%), the limbs and the bones (2%), and the mediastinum (1%).<sup>4</sup> They occur due to the lack of development of communication between the lymphatic and the venous systems during the sixth week of intrauterine life.<sup>2</sup>

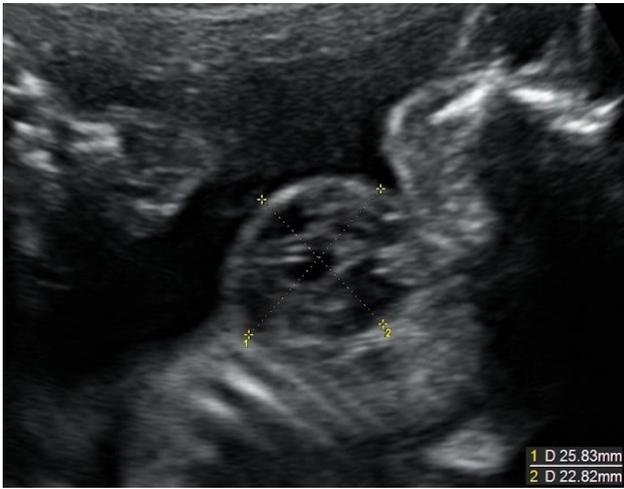
Unlike cystic hygroma, axillary lymphangioma appears not to be associated with fetal aneuploidy and is diagnosed later in pregnancy.<sup>1</sup> The diagnosis is based on ultrasonography, appearing as unilocular or multilocular cystic masses with no blood flow on the color Doppler.<sup>3</sup> Tumor growth, hydrops fetalis, fetal anemia secondary to intralesional bleeding and shoulder dystocia are possible obstetric complications of this malformation.<sup>1</sup> Surgical excision of the tumor and affected tissues is the main technique of choice and sclerotherapy is an alternative treatment.<sup>3</sup>

Therefore, prenatal diagnosis is essential to determine parents' counselling, plan the mode of delivery and the postnatal care. The authors report a case of a right fetal axillary lymphangioma and review the literature.

### CASE REPORT

A 38-years-old primigravida, with no relevant medical history, was diagnosed at 21 weeks' ultrasound with a fetal right axillary mass. The mass had a multilocular structure, thin septa and well circumscribed borders, without blood flow in color Doppler and it spreads from axilla to right anterior chest wall measuring 26×23 mm, without apparent infiltration of thoracic wall (Figure 1). The hypothesis of a lymphangioma was placed. No other fetal anomalies or makers of aneuploidies were noted at this exam. The first trimester ultrasound and aneuploidy screening have been normal. Invasive chromosome analysis was refused by the parents and after counseling the couple opted by expectant management. Fetal echocardiography was normal and serial ultrasounds revealed a gradual increasing in mass dimensions, reaching 98×62 mm at term (Figure 2), with no other alterations, namely hydrops, fetal malformations or

infiltration of adjacent organs by the lesion. The pregnancy was complicated by the diagnosis of gestational diabetes at 28 weeks, which was controlled with insulin and no other intercurrents were noticed.



**Figure 1: Second trimester ultrasound at 21 weeks of gestation, showing a right axillary mass consistent with lymphangioma.**



**Figure 2: Third trimester ultrasound at 31 weeks of gestation revealing an increase in mass dimensions.**



**Figure 3: Appearance of right axillary lymphangioma at birth.**

A cesarean delivery was performed at 39 weeks due to the mass dimensions and risk of labor obstruction. A male neonate weighing 4010 g, with an Apgar score of 10/10 was delivered. Neonatal examination revealed a soft cystic right axillary mass measuring 10 cm without other anomalies (Figure 3).

The infant was sent to a reference department of pediatrics surgery and underwent a magnetic resonance, which revealed an extra-thoracic multilocular mass with 105×55×110 mm, compatible with a lymphatic malformation. The infant had an uneventful surgical excision and the pathological exam confirmed the diagnosis of lymphangioma. At two years of age, the baby has a normal psychomotor development.

## DISCUSSION

The lymphatic channels are formed in the sixth week of intrauterine life which later communicates with the venous system. Failure to establish venous drainage is the basis of lymphangioma pathogenesis.<sup>2</sup> Lymphatic malformations vary in size from a few millimeters to several centimeters in diameter.<sup>5</sup> Lymphangiomas are histologically classified into three types: capillary lymphangiomas, comprising of lymphatic channels and likely to expand to involve adjacent tissues; cavernous lymphangiomas, consisting of dilated endothelial-lined lymphatic sinuses and cystic lymphangiomas made of various sizes of dilated lymphatic vessels. All types can coexist in a same lesion.<sup>3,6</sup>

Lymphangiomas are rare benign tumors, mostly located in the neck, also known as cystic hygroma. The axilla is the second most frequent location of this tumor.<sup>1,3</sup>

Prenatal diagnosis of lymphangiomas is made by ultrasound. Unlike cystic hygroma, axillary lymphangioma is typically diagnosed later in pregnancy, as in the present case where the malformation was noted at 21 weeks.<sup>7</sup> In the ultrasound lesion may be unilocular, but more often the structure has been multicystic, hypoechoogenic, with thin septa and may contain solid components.<sup>4,7</sup> The absence of blood flow on color Doppler mapping is characteristic of lymphangiomas.<sup>2</sup> The detection of blood flow signals within the mass by color Doppler raised the possibility of an associated hemangiomatous component or tumor bleeding.<sup>8</sup>

The differential diagnoses of this malformation comprise hemangioma, hemagio-lymphangiomas, teratomas, ectopia cordis, body stalk anomaly and limb body wall defect.<sup>9,10</sup> Additionally, fetal axillary lymphangiomas may be part of some genetic syndromes such as Parkes Weber syndrome, Maffucci syndrome, Proteus syndrome and Klippel-Trènaunay syndrome.<sup>9,10</sup>

When lymphangioma is diagnosed in the antenatal period, fetal karyotype may be considered to provide accurate diagnosis and genetic counselling. Distinct from

cystic hygroma that is associated with fetal aneuploidies, mainly Turner syndrome and trisomy, axillary lymphangioma appears not to be associated with chromosomal abnormalities.<sup>2,11</sup> In this case report parents refused amniocentesis and opted for expectant management.

Shoulder dystocia, fetal anemia secondary to intralesional bleeding, hydrops fetalis, fetal distress and stillbirth are possible obstetric complications of this condition.<sup>1,6</sup> The antepartum management and the mode of delivery should be individualized, since there is no standardized recommendation for prenatal management.<sup>4,9</sup> Ultrasound evaluation should be undertaken in order to detect fetal hydrops and anatomical anomalies. Repeated evaluations may be necessary for the evaluation of the tumor growth, since lymphangiomas may expand and infiltrate adjacent organs.<sup>7</sup> Magnetic resonance allows a better estimate of tumor extent and relation with contiguous structures.<sup>2,4</sup> In our case, we started the serial ultrasound at the time of diagnosis to monitor tumor growth, signs of hydrops or fetal abnormalities. When large lesions are present, a cesarean delivery may be advisable, as they can lead to obstructed labor and neonatal morbidity.<sup>7</sup>

Postnatal prognosis is generally promising in the absence of fetal hydrops or structural malformations, invasion of neighboring tissues and abnormal karyotyping.<sup>11</sup> Since these tumors do not resolve spontaneously after childbirth, surgical excision of the lesion and affected tissues are necessary. Infiltrative and extensive masses are difficult to be extracted surgically. If total surgical excision is not carried out, recurrence may be seen.<sup>5</sup> Sclerotherapy is considered appropriate for the treatment of the lesions which are not resectable surgically.<sup>3,5</sup>

Antenatal diagnosis of this condition permits a better anticipatory care – investigation of associated congenital anomalies, planning the mode of delivery and orientation of newborn to appropriate care.

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