Fetal axillary lymphangioma diagnosed on a 2D/4D ultrasound second trimester scan – a case report and short literature review

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Abstract
Fetal lymphangioma, also known as cystic hygroma, is a hamartomatous congenital tumor which involves the fetal skin and the subcutaneous tissue. The most common site of appearance is in the neck region. Location of the lesion in the axilla as well as in other anathomical sites is very rare.

Prenatal diagnosis can be made on ultrasound examination. Other structural or chromosomal anomalies are frequently associated with this diagnosis. The antepartum management and type of delivery should be set up by a multidisciplinary team.

We present a rare case diagnosed with axillary lymphangioma during the second trimester morphological ultrasound 2D/4D scan.

Keywords: lymphangioma; cystic lymphangioma; cystic hygroma; fetal vascular tumors

Introduction
Fetal lymphangioma is a hamartomatous congenital tumor which involves the fetal skin and the subcutaneous tissue. In most of the cases (70-80%) the tumour is located in the neck region and secondly in the axilla (20%). Other rare, uncommon, locations such as mucous membranes, mediastinum, pelvis, extremities, chest wall, retroperitoneum (kidneys) and intraabdominal area (colon, spleen, liver) have been reported [1-3]. A multidisciplinary team is mandatory to successfully manage the antepartum period as well as the delivery of the fetus and the further medical and/or surgical approach of the newborn.

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Case report
A 27 year old pregnant woman (gravida 1, para 1) was admitted for a second trimester morphological scan. All previous scans were within normal limits and there was no significant medical or gestational history. First trimester screening for chromosomal anomalies showed a low risk pregnancy. Also, no familial history of congenital anomalies was recorded.

Ultrasonographic evaluation revealed a 23 week and 1 day live fetus, with a left axillary mass. The mass was well-circumscribed, multilocular in structure, measuring 21/18 mm in two dimensions (fig 1a,b). No other structural abnormalities were noticed. There was no blood flow within the mass on color Doppler examination (fig 1c). In a short moment of upper limb abduction, we were able to 4D scan the axillary region which clearly revealed the mass (fig 1d).

According to the ultrasonographic characteristics of the axillary mass, we were able to raise the suspicion of a vascular axillary tumor, probably a lymphangioma due to the absence of blood flow on Doppler scan.
Genetic testing was recommended, which showed normal fetal karyotype, and counseling was offered. The pregnancy continued uneventfully. A female baby weighting 3250 grams, Apgar scores 8 and 9 at 1 and 5 minutes, was delivered by elective cesarean section at 38 weeks of gestation in another medical facility. First examination of the newborn revealed the left cystic soft axillary mass measuring 60/45 mm. The baby was admitted to a pediatric surgery unit where, after comprehensive evaluation, was operated on day 6 after birth. Pathological report of the lesion confirmed the diagnosis of axillary lymphangioma. The evolution was uneventful afterwards with no recurrences until 1 year of age.

Discussion

The malformation occurs when the embryonic lymphatic sacs fail to connect with the venous system [4]. There are reports that suggest that large multilocular, septated cysts results from the complete obstruction of the lymphatic sacs, whereas a nonseptated cyst is the consequence of temporary accumulation due to incomplete obstruction of lymphatic drainage [5,6].

The reported prevalence of fetal lymphangioma is 1.1-5.3 per 10,000 births and is dependent on maternal age, race and residence [7,8]. Both genders are equally affected [9].

It is known that some genetic disorders, such as Turner syndrome, Klippel-Trenaunay-Weber syndrome, Maffucci syndrome, CLOVES syndrome, Proteus syndrome, are associated with vascular malformations [10,11]. Yet, when lymphangioma is detected early in pregnancy and is located in the dorsal or nuchal region the risk of aneuploidy is even higher and strongly suggestive of trisomies 21, 18, 13, Turner or Noonan syndrome [9,12,13]. That is why the overall prognosis is poor. Only 9 % of cases are healthy children with normal karyotype [14].

Prenatal diagnosis is based on ultrasound examination and magnetic resonance imaging [15]. Fetal lymphangiomas typically appear on ultrasound as a multiloculated cystic, sonolucent, septated mass, possibly associated with hemorrhage areas within the cyst [16]. In addition, abnormalities of the ipsilateral limb, such as deformities or hypo/hypertrophies, are also reported [17]. If a lesion has compressive effects on important fetal vessels, it may cause fetal hydrops [3]. Generally, the cystic tumors progressively enlarge during pregnancy. Spontaneous regression is exceptional.

The distinctive feature of the lymphangioma and also the characteristic that helps distinguish it from a haemangioma, is the absence of blood flow. Color Doppler ultrasound reveals, as in our case, no venous or arterial blood flow within the mass [18].

Magnetic resonance imaging can provide information about the tumor’s infiltration in the thoracic structures [1]. A cytogenetic study for suspected aneuploidy should always be performed [19].

Antepartum management and the type of delivery should be individualized. There are no standardized recommendations. The postnatal management depends on the tumor’s size and its location [15]. Spontaneous regression is very rare, thus excision is required [9]. Unfortunately, local recurrence has been reported after surgery [20].

Prognosis is associated with the existence or not of the chromosomal anomalies, the penetration or invasion of the adjacent structures, hydrops fetalis. The size and septation of the mass do not predict the prognosis reliably [19].

Conclusions

The fetal vascular tumors are challenging clinical entities. Their prognosis can be good, as in our case, in the absence of fetal hydrops, invasion of the neighboring tissues, and abnormal karyotype. A multidisciplinary clinical team is always required in order to successfully manage such rare cases.
References