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Case Report

Obstetrics and Gynaecology Section

An Unusual Origin of Fetal Lymphangioma Filling Right Axilla

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ABSTRACT

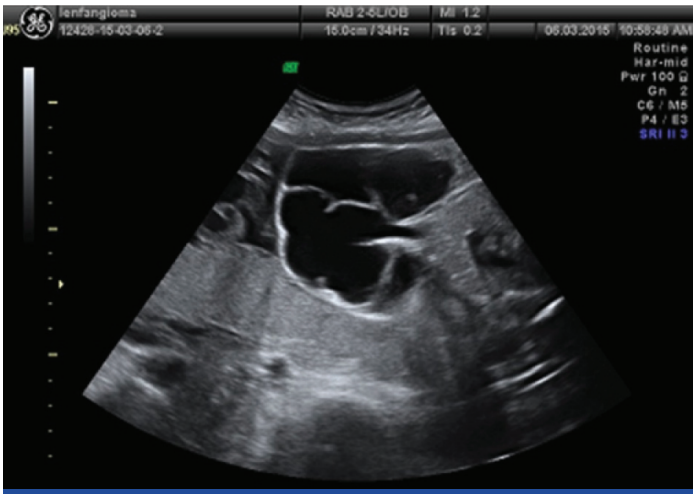
Fetal lymphangioma is a hamartomatous congenital anomaly of the lymphatic system, which is embracing the fetal skin (sometimes mucous membranes) and the subcutaneous tissue. The general consensus is that it occurs as a result of failure in lymphatic drainage. A 36-year-old pregnant woman was referred to our perinatology clinic at 22 weeks' gestation, because of a fetal right-sided axillary mass revealed by ultrasonography. The mass measuring 5x7x7cm in three dimensions had a multilocular structure without colour Doppler flow and well-circumscribed borders. Amniocentesis revealed a normal constitutional karyotyping. Lymphangioma was considered as prediagnosis. A healthy female baby weighing 3470 grams was delivered at term. Neonatal examination and the postnatal MRI confirmed the diagnosis. The baby is still on follow-up with the medical treatment of Sirolimus an anti-proliferative drug, and the mass got smaller significantly in 8 months after delivery.

Keywords: Axillary mass, Chest wall mass, Karyotyping, Magnetic resonance imaging

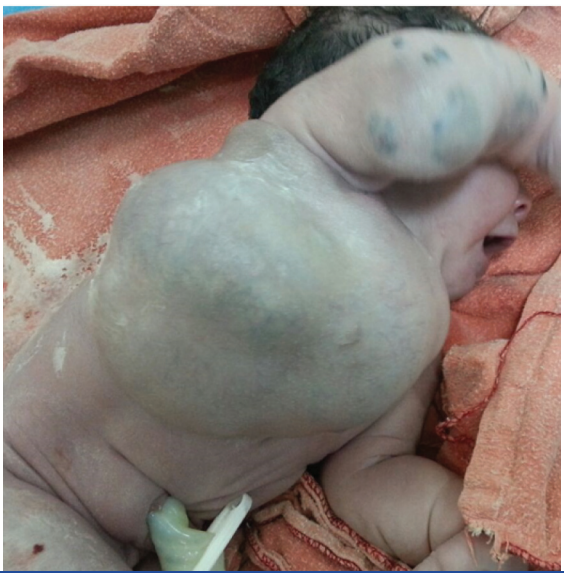
CASE REPORT

A 36-year-old multiparous (gravida 4, para 2, dilatation and curettage 1) pregnant woman who admitted to our perinatology outpatient clinic, because of a fetal right-sided septated axillar mass revealed by ultrasonographic investigation at 22nd week of gestation. The mass had a multilocular structure and well-circumscribed borders and had a measure that was 5x7x7cm in three dimensions. It was originating from the fetal chest wall, and filling the right fetal axilla. The colour Doppler ultrasonographic examination revealed no blood flow through the mass and the detailed fetal anatomic scan revealed no other anomaly. First and second trimester aneuploidy scans were all in normal ranges. The woman was in a non-consanguineous marriage and had no significant medical or gestational history. No familial history of congenital abnormalities was noted in maternal and paternal genetical inquiry. One of her previous two children died of mitochondrial encephalopathy at 13th years, the gender was female. The first child was a 20-year-old healthy male. After a detailed counseling, the family chose the expectant management until the delivery. Amniocentesis was performed and revealed a normal constitutional karyotyping. Thereafter, serial ultrasound studies demonstrated that the lesion increased in size slowly, it reached the size of 7x8x10cm at 28 week's gestation, 9x10x11cm at 32 weeks' gestation [Table/Fig-1]. In the follow-up ultrasonographic examinations, it was seen that there were areas of intracystic haemorrhage in some of the locules and the right arm of the fetus was in the position of abduction, continuously [video-1,2].

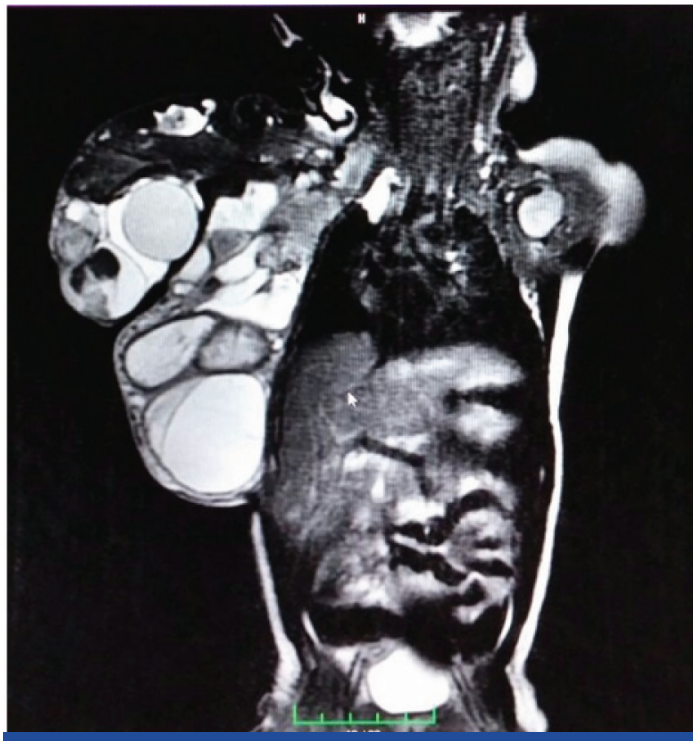
The fetus was delivered via cesarean section at 38 weeks' gestation, when the active labor started. Cesarean section was chosen to avoid possible fetal trauma during vaginal birth. A female baby weighing 3470 grams was delivered with the Apgar scores of 7 and 9 at 1 and 5 min, respectively, in the presence of an expert neonatologist. Neonatal examination revealed that the cystic mass (measured 10x12x14cm in three dimensions) covered the right anterior and lateral chest wall, and extended to the right upper arm through the axillary region. Also, it was related to a smaller cystic lymphangioma at the apex of the posterior axillary region, measured 3cm in three dimensions and also there were small various sites of skin discolouration and millimetric lymphangiomas



[Table/Fig-1]: Ultrasonographic view of the multilocular cystic mass filling the right fetal axilla.



[Table/Fig-2]: Postnatal view of the mass and minor skin discolourations on the ipsilateral upper extremity.



[Table/Fig-3]: T2-weighted magnetic resonance imaging of the mass and intense enhancement of the contrast material by the cyst walls and septa.

[Table/Fig-4]: The tension of the mass was eased, the covering skin loosened and folded significantly, in an 8 months of continuous treatment with Sirolimus.

on the ipsilateral upper extremity [Table/Fig-2]. Small lesions were defined as capillary lymphangioma. The neonatal vital signs were unremarkable at all.

The T2-weighted images on the MRI revealed that the mass was consistent with cystic lymphangioma [Table/Fig-3], also cyst walls, and septa showed intense enhancement after the administration of council material, and there were areas of parenchymal consolidation in both lungs located in postero-basal segments. There was no sign of pulmonary failure.

A conseil of paediatric surgery and oncology reached a consensus that surgery might lead to a huge defect on thoracic wall, and medical treatment with oral Sirolimus (an anti-proliferative and immunosupressive drug that effects on lymphatic cell lines) was the chosen treatment modality. In an 8 months of continuous treatment, the tension of the mass was eased, the covering skin loosened and folded significantly [Table/Fig-4]. The treatment with Sirolimus was found as successful and it was decided to continue. The baby is still being followed by paediatric oncology team with hope for complete resolution.

DISCUSSION

Fetal lymphangioma is a hamartomatous congenital anomaly of the lymphatic system, which is embracing the fetal skin (sometimes mucous membranes) and the subcutaneous tissue. The general consensus is that it occurs as a result of failure in lymphatic drainage. In general, it is detected during routine prenatal ultrasonographic survey. It has generally thin walled, multilocular structure. It is reported to be found in the fetal neck mostly (75%), and secondly in the axilla (20%). There are also uncommon locations, like mucous membranes, mediastinum, pelvis, extremities, chest wall, and intraabdominal area. Detection of it in uncommon locations usually occur in the second half of the pregnancy. The distinctive feature of the lymphangioma is the absence of blood flow in it with the use of colour Doppler ultrasonography in order to distinguish it from haemangioma [1].

Two types of lymphangiomas have been described according to the depth and size of the lesions: Lymphangioma circumscriptum (the small and superficial one), and cavernous lymphangioma (the bigger and deeper one). Cystic hygroma is generally accepted as a sub-type of cavernous lymphangiomas [2].

Our case is about a progressive enlarging fetal cystic lymphangioma throughout the pregnancy, and getting smaller with the use of Sirolimus during and after the neonatal period. This case was located at the chest wall extruding to right fetal axilla. The cystic lymphangiomas have been seldomly reported to be located at the chest wall [1,3]. Also, it was related to a smaller cystic lymphangioma, various ipsilateral sites of skin discolouration, and millimetric lymphangiomas.

The fetus did not have any other skeletal, vascular and lymphatic abnormalities, so the presence of Klippel-Trenaunay-Weber syndrome (also called angioosteohypertrophy syndrome, haem-angiectatic hypertrophy or osteohypertrophic naevus flammeus) associated with hemihypertrophy seemed unlikely [4].

It has been believed that a septated fetal lymphangioma takes its origin from the failure of drainage of the lymphatic ducts into the final venous vessels. Its prevalence has been reported to be about 1.1-5.3 per ten thousands births [3], and has been reported as not related to fetal gender [4]. To a large extent, it is septated. However, a nonseptated lymphangioma is thought to be sourced from a temporary obstruction of the lymphatic drainage [5]. The fetal lymphangiomas generally appear on ultrasound as multicystic, sonolucent, septated masses which are prone to increase in size during gestation. Colour Doppler ultrasound reveals no venous or arterial blood flow within the mass. Ipsilateral extremities are prone to have additional abnormalities like limb reduction deformities, hemihypertrophy [4]. The prognosis has been reported to be associated with the penetration or invasion of the neighbouring tissues. The size and septation of the mass do not predict the prognosis reliably [6]. If a lesion has compressive effects on important fetal vessels, it may cause fetal hydrops [7].

The risk of aneuploidy is reported to be higher, if the lymphangioma located in dorsal or nuchal region [8]. Also, it was stated that fetal lymphangioma detected in early gestation has a more inclination to be associated with aneuploidy [9].

The chest wall lymphangioma has specifications which distinguish it from other locations. Its differential features include: Its location is rarely seen; it is usually seen later in gestation; and it has a rare association with aneuploidies in comparison with the lesions in other locations.

The fetal MRI would provide more information about the infiltration of the tumour to the fetal thoracal structures, but we could not deny the superiority of the 2D gray scale ultrasonographic images. The postnatal MRI provided useful results about the detailed structure and neighbouring relationships of the mass. The fetal MRI has been found as the most useful diagnostic device in case of fetal neck masses, considering the potential necessity of the Ex-utero Intrapartum Treatment (EXIT) procedure [10]. In previous reports, fetal lymphangiomas have been described in various locations like neck [5], axillary region, tongue [11], cervicothoracic region [12], and other sites including thoracic wall [7]. Our case is similar to other thoracoaxillary lymphangiomas [1,13], but the present medical treatment with Sirolimus is unprecedented in thoracic wall cases, according to the literature.

Although the treatment of surgical excision is conceived in the foreground, the choice may change by patient to patient or experience of the clinician [9]. In a study by Grabb et al., it was showed that there have been complete involution in the next 5 years follow-up [14]. Local recurrence has been reported after surgery.

Sclerosing agents like Bleomycin and OK-432 (inactivated streptococcal organisms) can be injected directly into the mass for achieving the shrinkage totally, or if not, partially prior to surgery [15]. The administration of OK-432 has been reported as successful [4]. Also, there is a case report in the literature which reported absolute cure of fetal cystic hygroma with the use of OK-432 injection via the abdomen of the pregnant woman [16]. Tanigawa et al., reported that 39% of the lesions got shrunk with the use of bleomycin which is known as a fibrotic agent. Its use was recommended especially for recurrent lymphangioma cases after unavoidable incomplete resection [17].

Propranolol inhibits VEGF and promotes apoptosis on lymphangioma cells [18]. Generally, its use has been mentioned as safe for paediatric patients, but there are some doubts about its efficacy [19].

Sirolimus is an anti-proliferative and immunosupressive drug that effects on lymphatic cell lines. It has been reported as the inhibitor of the VEGF receptor-3, also an inhibitor of lymphatic endothelial generation [20]. It made the lesion smaller significantly, and was defined as a successful therapy for this case.

CONCLUSION

The management of cystic lymphangiomas is a challenging clinical entity. However, the prognosis is generally promising with the absence of fetal hydrops, invasion of the neighbouring tissues, and abnormal karyotyping. Medical approach may outweigh surgical procedures, like in our case. Multidisciplinary clinical approach including expert perinatologists, paediatric surgeons, and neonatologists is a necessity to manage such a rare case successfully.

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