The value of early recognition of fetal lymphangioma

Abstract

Introduction: Lymphangiomas are very rare benign tumors of the lymphatic system, appearing as uniseptate or multiseptate cystic masses, which are usually located in the cervical or axillary area. Postnatal outcome depends on the size and location of the lesion. An increasing number of such congenital abnormalities are detected on routine conventional prenatal ultrasonography. Although prenatal evaluation for the prognosis of fetal lymphangioma has been based on two-dimensional ultrasonography, magnetic resonance imaging may help in assessing the extent of a lesion. Isolated lymphangiomas generally have a favourable prognosis and sclerotherapy or surgical resection is effective in most of the cases.

Case presentation: We present two cases of fetal axillary lymphangioma. In the first case, the lymphangioma was diagnosed antenatally, so parents were comprehensively counselled and post natal follow up was organised at a tertiary hospital. The second case remained undiagnosed until birth. This caused significant distress to both the parents and clinicians, especially after a coincident traumatic delivery.

Conclusion: Both these cases emphasise that early diagnosis of fetal lymphangioma is critical and a combination of ultrasonography and magnetic resonance imaging can facilitate detection of lesions which are relatively limited and accessible to therapy in utero. This would also enable clinicians to perform a karyotype and comprehensively consult parents regarding the treatment and delivery options as well as outcome of the pregnancy.

Keywords: cystic hygroma, fetal lymphangioma, lymphangioma.

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Introduction

Lymphangiomas are hamartomas of the lymphatic vessels which may infiltrate surrounding structures. They occur roughly in one in every 6000 live births.[1] About 50% are present at birth and up to 90% become evident by two years of age.[2] These are benign tumors of the lymphatic system, appearing as uniseptate or multiseptate cystic masses. Lymphangiomas are frequently located in the area of the neck in 75% of cases (cystic hygroma). Uncommonly, they may be present in the axilla in 20% of cases.[2] These benign fluid-filled cystic masses may result in displacement of adjacent structures. A small percentage of these lesions may present as giant neck masses. Lymphangiomas may be macrocystic, microcystic or mixed lesions. Location, extension of the lesion and other associated abnormalities determine the prognosis of a lymphangioma. Since fetal lymphangiomas are usually associated with karyotypic or other abnormalities, such as polyhydramnios and hydrops fetalis, their prenatal diagnosis is essential. The neonatal outcome of a large fetal lymphangioma is generally poor.[3] A number of such congenital abnormalities are identified on routine prenatal ultrasonography (USS).[4–6] The absence of blood flow on color Doppler mapping is characteristic of lymphangiomas compared to hemangiomas.[7] Prenatal assessment for the prognosis of fetal lymphangiomas has been based on two-dimensional ultrasonography (2DUS).[5,6] However, Magnetic resonance imaging (MRI) may be helpful in assessing the extent of a lesion.[8] While spontaneous regression can occur in a fetal lymphangioma with normal chromosomes, large fetal lesions necessitate a perinatal multi disciplinary team approach. This facilitates a discussion of potential management options, including prenatal cyst aspiration, and planning a mode of delivery that will avoid fetal damage.[9] Therefore, prenatal diagnosis allows a planned delivery, and adequate intrapartum and postnatal resuscitation may improve the prognosis.

Case presentation

A 30-year-old primi gravida woman was referred to antenatal clinic from emergency department at 15 weeks of gestation for follow up after an ante-partum hemorrhage. Morphology scan, at 20 weeks although suboptimal, failed to show any fetal abnormalities. A second USS reassessment 23 weeks revealed a 45 x 18 x 38 mm multi-loculated cystic structure on the right chest wall, extending

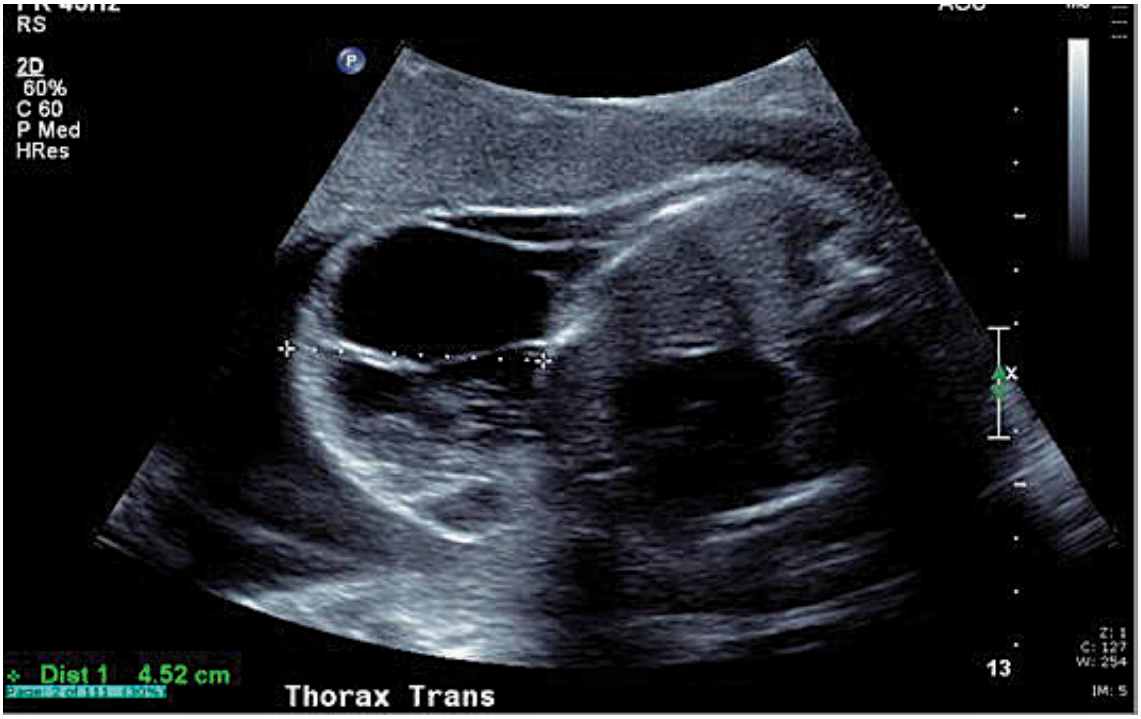

Figure 1: Demonstrates significant growth of the multi-loculated lesion on the right chest wall, measuring 74 x 39 x 55 mm.

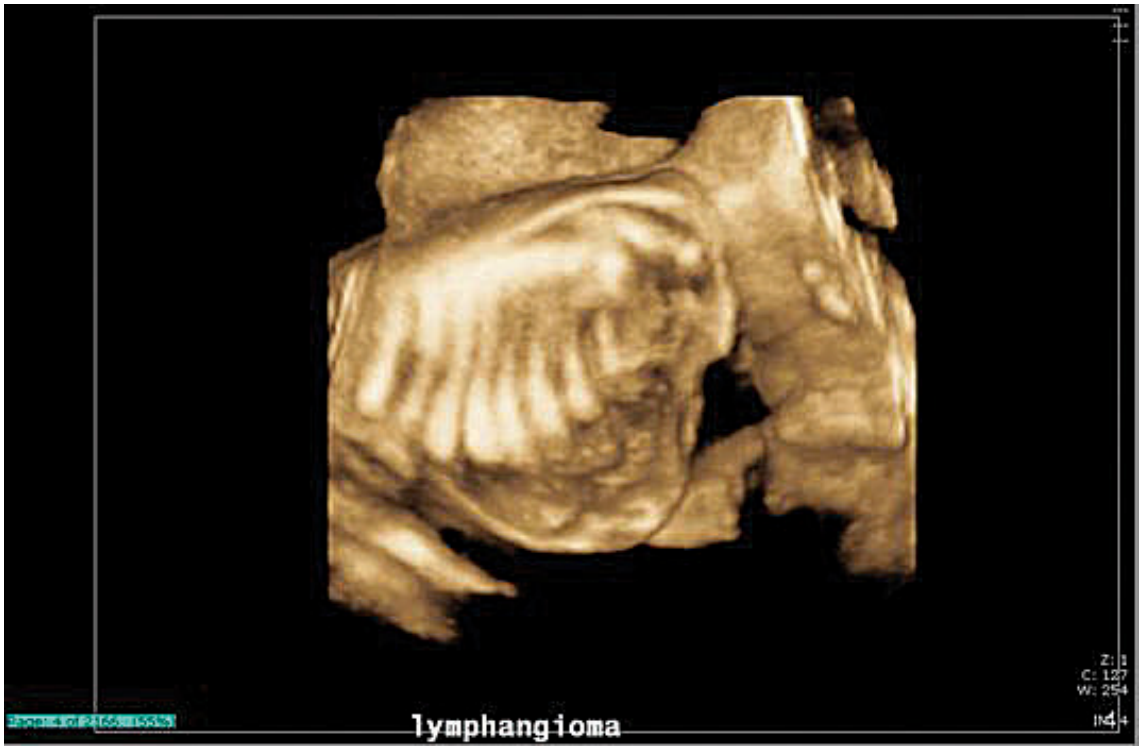

Figure 2: 3D image at 29 weeks, showing that the Lymphangioma is stable in its size.

from the axilla to the mid abdomen. It did not contain color flow on Doppler and was suggestive of a lymphangioma. On follow up USS at 27 weeks, the lymphangioma had grown significantly and measured 74 x 39 x 55 mm (Figure 1). It remained stable at the 29 weeks scan (Figure 2).

The result of an amniocentesis performed at 23 weeks was consistent with a normal karyotype. The patient was referred to a tertiary centre for second opinion USS and MRI evaluation. Multi–planar T1 and T2 HASTE images acquired through the fetal chest and abdomen revealed a large cystic axillary mass that was closely associated with the right lateral chest wall and axilla without an intra thoracic or intra abdominal component. In addition, there was a tail of the mass extending deep into the axilla and it was likely to be intricately related to the axillary

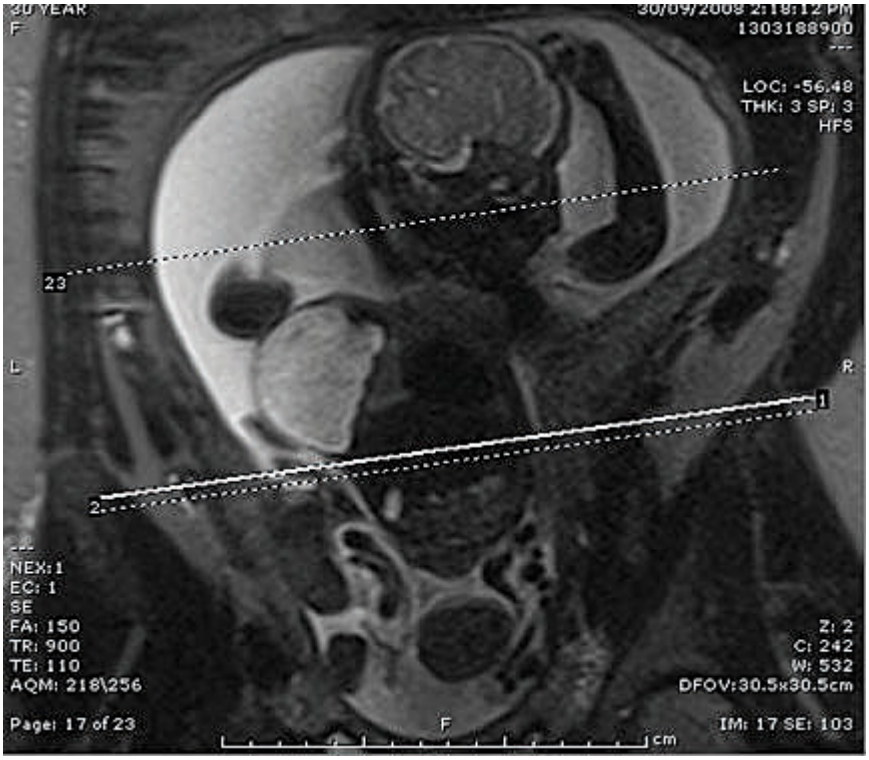

Figure 3: Fetal MRI at 25 weeks, confirming no intra thoracic or intra abdominal extension.

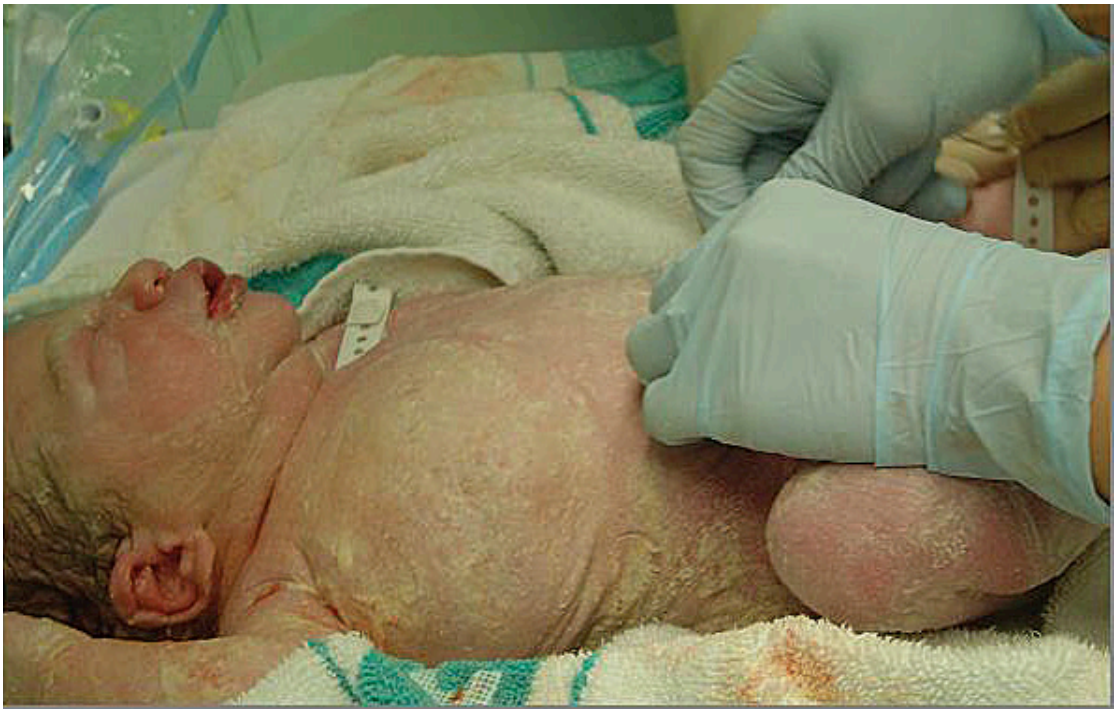

Figure 4: Baby at birth.

neurovascular bundle (Figure 3).

These features were consistent with USS findings and suggested the most likely diagnosis of a lymphangioma. The multi-disciplinary team extensively counseled the patient, including advice from an obstetrician and a paediatric surgeon regarding the mode of delivery and further management. Follow up USS were performed until 35 weeks of gestation, which indicated that the mass was stable in size. The baby was born by an uneventful caesarean section at 39 weeks (Figure 4).

Postnatally, Sclerotherapy performed at six months of age was unsuccessful. Therefore, the baby underwent a surgical excision at 22 months, which was complicated with fluid collection and infection, requiring a second procedure a few months later (two years). The post-operative course was straightforward. Histopathology confirmed a vascular malformation consistent with Lymphangioma (Figures 5a and 5b).

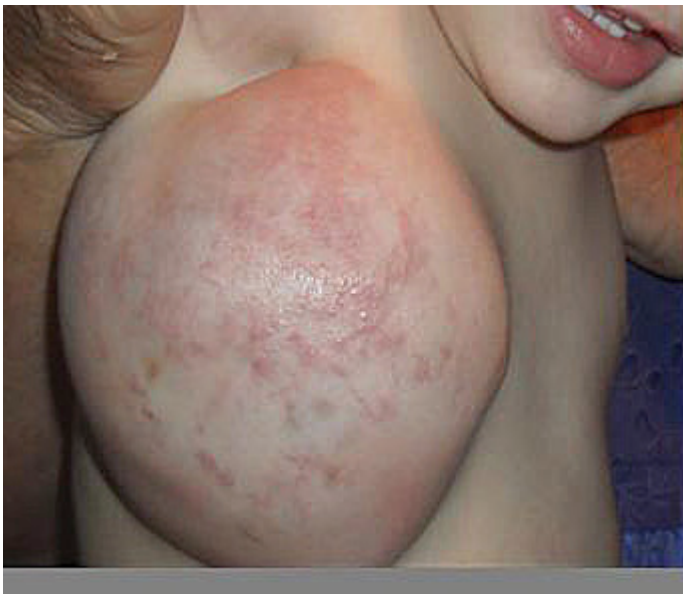

Figure 5a: Baby at 22 months.

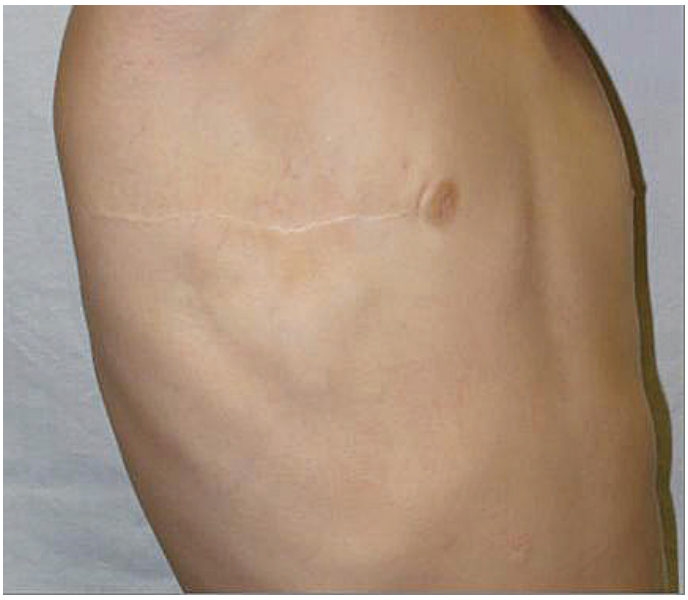

Figure 5b: Child at 4 years, post surgical excision.

Case 2

The second case is of an axillary lymphangioma diagnosed at birth in a 23-year-old healthy primi-parous lady who had four previous miscarriages. The antenatal course was complicated with several episodes of first trimester bleeding. The morphology scan and a third trimester scan both failed to diagnose the lymphangioma antenatally. The baby was born at 41 week gestation by instrumental delivery due to failure to progress in the second stage of labour. The baby weighed 4090 g and was admitted to special care nursery (SCN) for intravenous antibiotics and oxygen treatment.

A small right axillary mass was noted postnatally extending to the right nipple and neck. The mass was lobulated in areas and mobile. Trachea was clinically midline and not deviated. USS showed a multi loculated mass, measuring 34 x 42 x 20 mm within the superficial chest wall, extending from the level of the subclavian vessels, inferiorly to the mid axillary line and medially to the mid clavicular line (Figure 6). These findings were consistent with an axillary lymphangioma. The baby was stable and discharged from SCN after two days with a referral to vascular surgery clinic.

Though the pregnancy and delivery were not complicated by the lymphangioma, the diagnosis after a traumatic birth and need for an early operation was very stressful for parents, requiring further consultation and debriefing.

This further highlights the importance of ante natal diagnosis and planned delivery.

Discussion

Lymphangiomas are benign hamartomas of the lymphatic system, consisting of multiple dilated vessels. They occur because of a developmental defect in the lymphatic pathways, which usually develop from the sixth week of gestation, leading to the proximal dilatation of afferent channels. Lymphangiomas are histologically classified as: simple lymphangiomas, comprising of lymphatic capillaries, cavernous lymphangiomas, consisting of larger lymphatic vessels with a fibrous adventitia and cystic lymphangiomas or cystic hygromas, made of multiple cysts. All types can coexist in a same lesion. These cysts usually contain serous or chylous fluid, which may be complicated by bloody or purulent contents.[8] Almost 75% of lesions are located in the head and neck, or axilla, however the other 25% occur mostly in the extremities and trunk, and rarely in the mediastinum, abdomen and genitalia.[9]

The overall prognosis of a fetal lymphangioma is poor, with a mortality rate of 50 to 100%.[10] Karyotypic abnormalities and different malformation syndromes are present in 50 to 80% of patients with cystic hygromas. A study showed abnormal karyotypes in 69% of affected fetuses, the majority being 45XO,[11] but these features may not apply to non-nuchal lymphangiomas. Furthermore, fetuses with lymphangiomas secondary to a chromosomal abnormality may have additional health issues dependent on the underlying diagnosis.

At USS and MRI, cystic lymphangiomas are unilocular or multilocular cystic masses, with thin- or thick-walled septa. At USS, the fluid may be anechoic or there may be variable internal echoes or fluid-fluid levels, due to bleeding and fibrin deposition.[12] In this case, USS and MRI precisely identified the lymphangioma.

Historically, USS has been the imaging choice for prenatal assessment of fetal abnormalities due to its accuracy and safety, ease of use as well as low cost and availability.[13] However, more recently, ultrafast MRI has become a practical imaging tool for prenatal diagnosis, predominantly in patients with complex fetal abnormalities. Moreover, in this case, MRI permitted more comprehensive imaging of the fetus, and accurately defined the size of the mass.

In a published report of three cases, the authors described a case of fetal abdominal lymphangioma extending to an extremity diagnosed by USS.[3] Pregnancy was terminated in all these cases, considering the poor prognosis. In another case series, postnatal surgical resection or sclerotherapy was successfully performed in four patients with fetal abdominal lymphangiomas.[3]

Patients with an isolated cystic hygromas do have a very good

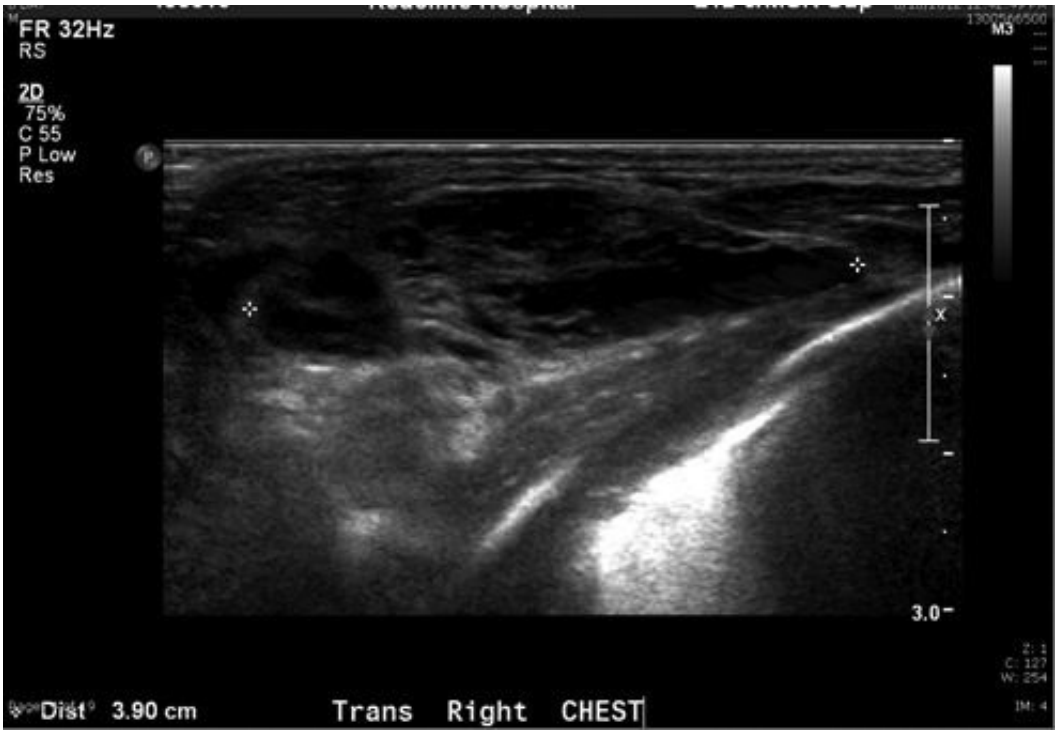

Figure 6: A multi loculated mass, measuring 34 x 42 x 20 mm within the superficial chest wall, extending from the level of the subclavian vessels, inferiorly to the mid axillary line and medially to the mid clavicular line

prognosis and surgical resection or sclerotherapy is efficient in the most of the cases. However, if the lymphangioma involves the tongue, floor of the mouth or airway structures, total resection is unlikely.

The preferred treatment for abdominal lymphangioma is surgical extirpation, with careful preservation of involved structures. Although the surgical treatment of diffuse and multiple lesions is exceptionally difficult and is associated with high morbidity and mortality, large but localised lymphangiomas can be excised completely.[10]

Sclerotherapy is considered appropriate for the treatment of those lesions which are not resectable surgically. Intralesional bleomycin, sclerotherapy with OK-432, or percutaneous embolisation with Ethibloc have all been reported to be effective.[3,14] Successful intrauterine treatment of a cystic hygroma with OK-432 has also been reported.[15]

Delivery may be an issue in patients with large lesions. A published case report has described a fetal death following shoulder dystocia and uterine rupture caused by a large axillary lymphangioma.[16]

In conclusion, these two cases highlight that early diagnosis of fetal lymphangioma is critical. A combination of ultrasonography and magnetic resonance imaging can identify lesions which may be accessible to therapy in utero. This also allows karyotyping to exclude associated chromosomal abnormalities and provides an opportunity to comprehensively consult patients regarding the treatment options, prognosis and the mode of delivery.

Consent

Verbal informed consent was obtained from both patients for publication and any accompanying images.

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

AK contributed to the interpretation of Ultrasound scans. AK and SF compiled and approved the final manuscript and ultrasound images.

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