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CASE REPORT

Axillary Cystic Hygroma : A Report of Two Cases

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ABSTRACT

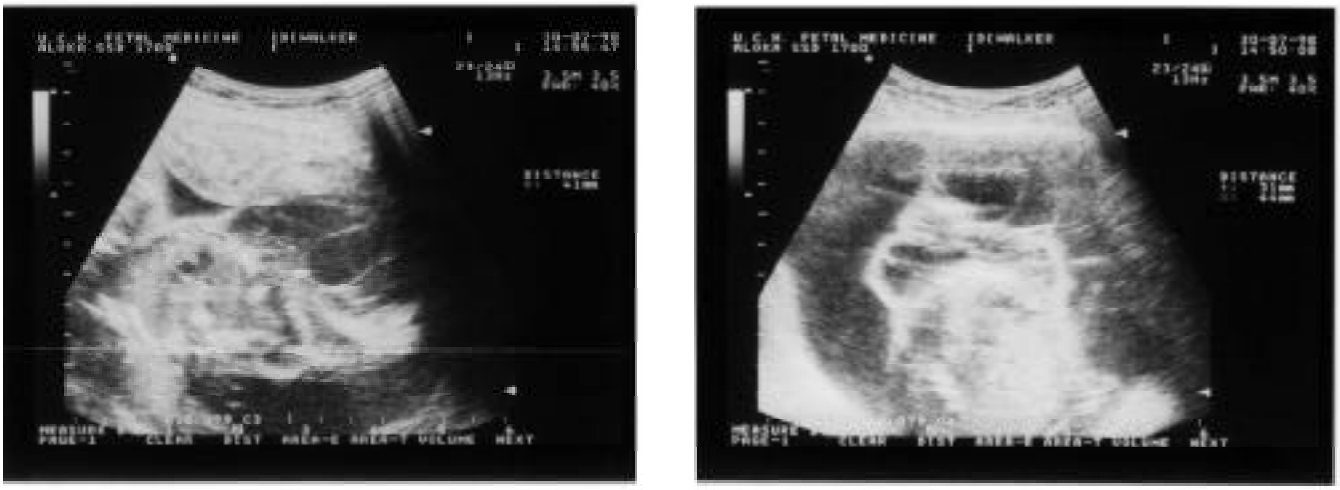
Cystic hygromas arising outside of the cervicofacial, thoracic and abdominal areas are extremely rare. We report two fetuses that were found at 20 weeks of gestation to have axillary cystic hygromas. Both had normal karyotype. Serial ultrasound showed progressive enlargement of the lesions without other abnormalities. Both the women underwent caesarean delivery and both neonates have undergone treatment as infants.

First reports of prenatal diagnosis of fetal cystic hygromas appeared more than 20 years ago.[1] These lesions may occur as isolated defects but are often manifestations of a variety of genetic syndromes or chromosomal abnormalities. Cystic hygromas probably develop from a defect in the formation of the lymphatic vessels which drain into two large sacs lateral to the jugular veins. As a consequence of this failure in connection, the jugular lymph sacs enlarge and lymph accumulates in tissues.[2] Fetal nuchal cystic hygromas however, are thought to be etiologically distinct from other cystic lymphangiomas which can occur in different sites including the axilla, mediastinum, mesentery and limbs.[3] These probably develop as a result of abnormal lymphatic growth, never achieving sufficient anastomoses with the larger lymphatic channels.[4] As a result of the different pathophysiology, prognosis and management are different from nuchal cystic hygromas.[5] We report two cases of axillary cystic hygroma diagnosed at the time of the routine 20 week anomaly scan.

Case Report I

A 28-year-old white woman, in her third pregnancy, was referred to the Fetal Medicine Unit at 20 weeks' gestation for a detailed scan following the detection of bilateral choroid plexus cysts during a routine anomaly scan. Both previous pregnancies had been uncomplicated, resulting in vaginal delivery at term. There was no relevance of past or medical family history. The scan performed at our Unit confirmed the bilateral choroid plexus cysts and in addition revealed an echogenic focus in the left ventricle and a multilocular cystic swelling in the left axilla, extending from the side of the neck to the mid thorax anteriorly, involving the left upper arm. The cystic swelling measured 4.1 x 3.1 x 4.4 cm. (Fig. 1a and b *–transverse and longitudinal*). All fetal biometry

measurements were consistent with the gestational age. The amniotic fluid volume and the placenta were normal. Both arms were of equivalent size. Colour Doppler imaging showed minimal blood flow through the lesion, which was not highly vascular in appearance.



a b

Fig. 1. a,b. The cystic swelling, measuring 4.1 X 3.1 X 4.4 cm. are presented in transverse and longitudinal section, respectively.

The differential diagnosis included haemangioma or cystic hygroma, although a combination of both could not be excluded. In view of the presence of the other soft tissue markers, an increased risk of aneuploidy was discussed. The parents decided to proceed with an amniocentesis, which was performed without complications. This showed a normal karyotype (46, XX). Serial ultrasound studies were performed every 3-5 weeks showing a normal fetal growth. The choroid plexus cysts disappeared at 27 weeks as did the echogenic focus, while the cystic structure involving the left axilla, upper arm and chest wall increased in size.

At 34 weeks', the cystic swelling extended around the humerus into the axilla and down the side of the thorax with its maximum diameter of 10x10 cm. around the upper humerus. In view of the size of the lesion and the potential risk of dystocia, fetal trauma and nerve damage, a caesarean section was performed at 37 weeks' of gestation. A 3350 grams female infant was delivered with Apgar scores of 7 and 8 at 1 and 5 minutes respectively. There was a large hygroma involving the left upper arm and axilla extending into the chest. Surgical excision of the cyst was performed without complication. The left upper extremity moved normally after the operation.

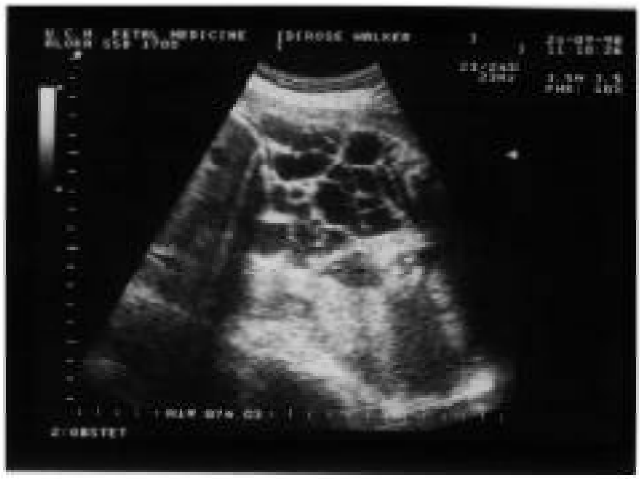
Case Report II:

A 32-year-old white woman, in her third pregnancy, was referred at 20 weeks to the Fetal Medicine Unit following ultrasound detection of a cystic mass surrounding the left humerus. Her two previous pregnancies had been uneventful, both resulting in vaginal deliveries of normal term infants. She was a non-smoker and there was no relevant past medical or family history.

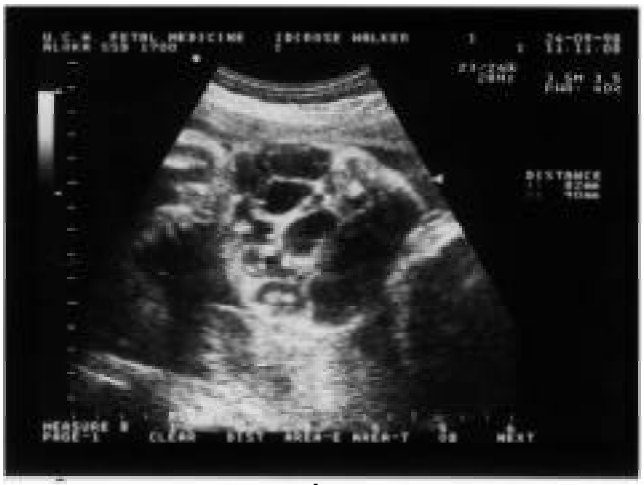
The ultrasound scan in Fetal Medicine Unit confirmed the presence of a multicystic mass surrounding the left humerus extending to the axilla and antero-lateral chest wall. The lesion measured

25 x 15 x 33mm and there did not appear to be any intrathoracic extension. Colour Doppler imaging did not demonstrate any flow in the lesion. No other abnormalities were seen. Placenta and amniotic fluid were normal. Both humerii were of equivalent lengths.

The parents were advised that the most likely diagnosis was a cystic hygroma, and haemangiomatous component could not be excluded. The woman had serial ultrasound examinations which were performed every 1-4 weeks. The fetus grew normally with normal amniotic fluid volume. The cystic lesion around the upper arm increased significantly in size. At 35 weeks, the appearance of the cystic swelling was unchanged, measuring 9 x 8 cm in cross section at the mid-humeral level (Fig 2 a and b). There was some involvement of the chest wall with minimal blood flow within the lesion. The arm was seen to move normally during the scan and both humerii remained of the similar length.



a



b

Fig. 2. a,b. The cystic swelling, measuring 9 X 8 cm in cross section.

In view of the risk of dystocia and fetal trauma a caesarean section was performed at 37 weeks. A 3289 grams male was delivered with Apgar scores 9 and 10 at 1 and 5 minutes respectively. There was a large cystic hygroma around the left upper arm and anterior chest wall with a haemangiomatous component to it. An ultrasound scan showed a large cystic hygroma, with small pockets of haemangiomatous areas within it. Surgical excision of the cyst was performed without complication. Pathological report showed haemaggiomatous component inside the cyst. The baby improved well after operation.

Discussion

Cystic hygromas can occur in a variety of anatomic locations. Detection in early pregnancy is quite common (around 1/120 pregnancies)[6] but the birth prevalence is only about 1/6000 deliveries, which is consistent with the fact that there is a high association with aneuploidy and many affected pregnancies are lost spontaneously. Furthermore a few lesions may regress spontaneously.[7]

There are two types of cystic hygromas, each with its own implications for chromosomal abnormalities. The first is located at the back of the fetal neck; it is associated with an increased nuchal

fold measurement and it usually contains internal septations. It can be seen in the first or in the second trimester and is associated with autosomal trisomies (i.e trisomy 21).[8,9] The prognosis is often poor.[10,11] The second type of cystic hygroma (cystic lymphangioma) is usually unilateral. It is usually located in the neck, axilla or abdominal wall, and is more commonly diagnosed late in pregnancy. This form is not associated with an increased incidence of chromosomal abnormalities.[9,12] The prognosis is good if other malformations are excluded.[13,14]

Axillary lymphangiomas account for appropriately 10% of all lymphangiomas.[15] They may be detected as a sonographically in the second or third trimesters when they often appear as small, transient, non-loculated cysts. A significant association (60%) with aneuploidy has been reported by one group,[13] but this has not been confirmed by others. Here we report two cases with serial ultrasonographic evaluations demonstrating an increase in size of these rare axillary hygromas.

However, there was a report that the use of magnetic resonance imaging (MRI) for the diagnosis of a cystic hygroma provides more information, give an accurate diagnosis; and can differentiate from the other similar ultrasonographic findings; which can not be differentiated by ultrasound such as the huge neck teratoma and cystic hygroma.[16]

The differential diagnosis in these cases included cystic hygromata, haemangiomatous lesion and a combination of both. As blood flow in haemangiomatous lesion may be minimal, colour flow Doppler investigations do not always exclude the presence of a haemangiomatous component. Counselling of parents can be difficult in these cases as postnatal management and prognosis will vary depending on the final diagnosis.

Fortunately both the pregnancies reported here continued normally without complication, although the axillary lesions increased in size significantly. There is good evidence to suggest that fetal dystocia is more likely to occur in the presence of large cystic lesions in the axilla area.[17,18] In order to avoid the risk of fetal brachial plexus damage in the cases we report here, caesarean sections were performed.[19]

Postnatal treatment of choice in these cases is surgical excision.[20] The prognosis after surgery is good with 10-15% of recurrence rate; which is depended on the surgical technique (whether completely or partial removal of the lesions),[21] the location of the lesion and the effect on the adjacent structures. Cases of spontaneous regression after birth are also reported.[22]

Medical therapy may be the best option for the treatment of a small cystic lesion or the residual lesions after operation. Bleomycin injection was demonstrated to be effective in the treatment of the residual lesions after surgery as well as fat emulsion into lymphangiomas which appear to be particularly suitable for patients with recurrence from partially removed lesions.[23,24]

In conclusion, we have reported two cases of axillary hygromata. Both infants have done well despite the rapid growth of the lesions prenatally. In both cases the prenatal diagnosis was accurate, although we consider that it is not possible to definitively exclude the possibility of a haemangiomatous component prenatally. The severe complication of axillary cystic hygroma is shoulder dystocia if a definite diagnosis is not obtained prenatally.

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