MMJ

CASE REPORT

Prenatal diagnosis of retroperitoneal lymphangioma – a case report

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Retroperitoneal lymphangiomas are rare congenital anomalies of the lymphatic system. Here we report a case of foetal retroperitoneal lymphangioma accurately diagnosed in the third trimester of pregnancy in a primigravida lady, using a combination of ultrasonography and magnetic resonance imaging. Prenatal diagnosis and multidisciplinary team approach is crucial in the management and prognosis of infants with lymphangiomas.

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Introduction

Lymphangiomas are congenital anomalies of the lymphatic system. These are benign fluid-filled tumours of the lymphatic vessels that may infiltrate or cause displacement of adjacent structures. Lymphangiomas are known to affect about 1 in 6000 live births, with approximately 50% presenting at birth and 90% becoming apparent at two years of age.[1] Retroperitoneal lymphangiomas are rare, accounting for only 1% of cases.[2] Foetal lymphangiomas may be associated with other problems and their prognosis depends on the location, size and the presence of other associated abnormalities. Here, we report a case of foetal retroperitoneal lymphangioma diagnosed in the third trimester of pregnancy.

Case Report

A 35-year-old Maltese lady, who was in her first pregnancy, was noted that her foetus had a large multiseptated right sided abdominal mass on routine visit at 33 weeks of gestation. (Figure 1). No other anomalies were detected. Previous ultrasound scans including a detailed anomaly scan at 21 weeks and a 4D scan at 28 weeks of gestation were normal. For further characterisation of the mass, a magnetic resonance (MR) imaging of the pelvis was carried out two weeks later and the diagnosis of a retroperitoneal lymphangioma was confirmed (Figure 2).

Figure 1 Multiseptated hypoechogenic avascular abdominal mass measuring 5.36cm by 4.55cm at the level of the lumbar spine on ultrasonography.

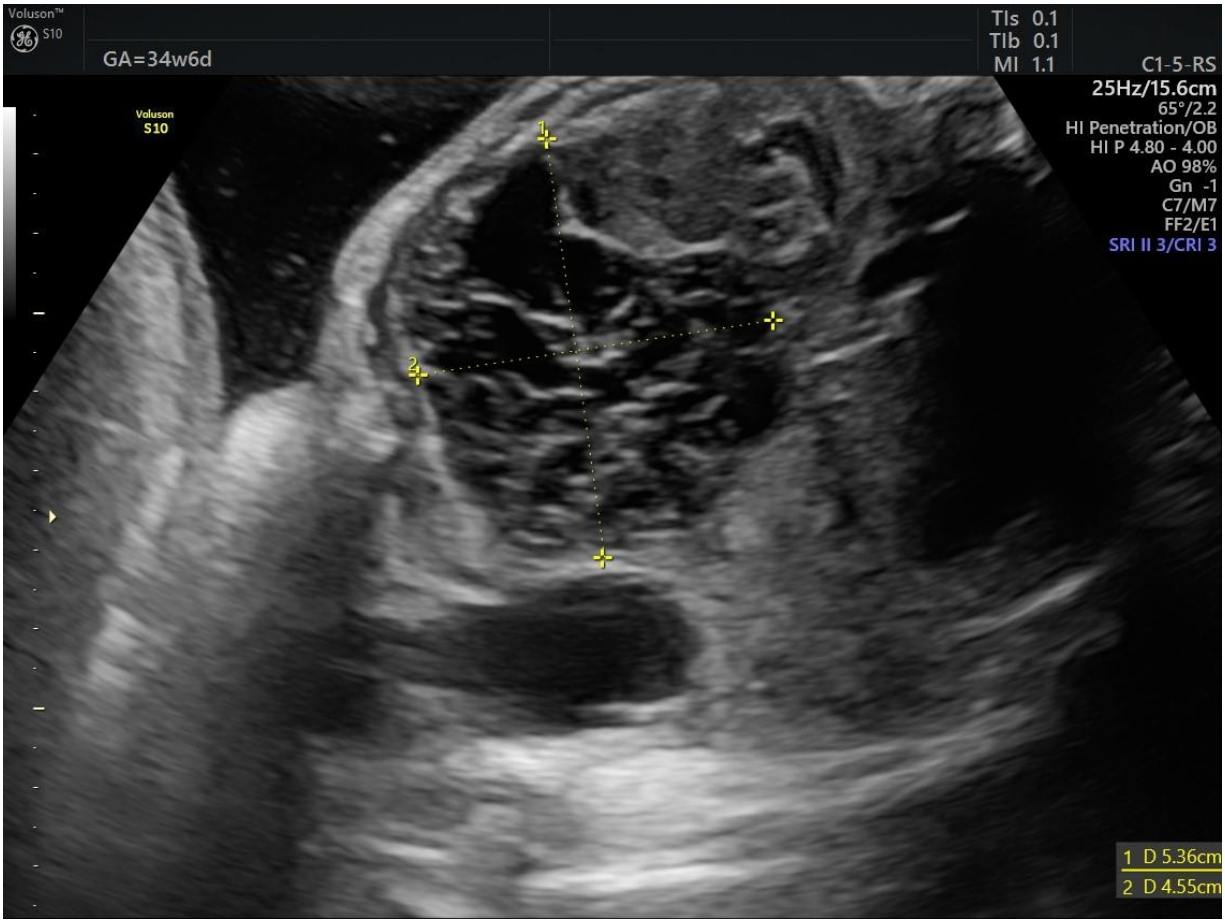
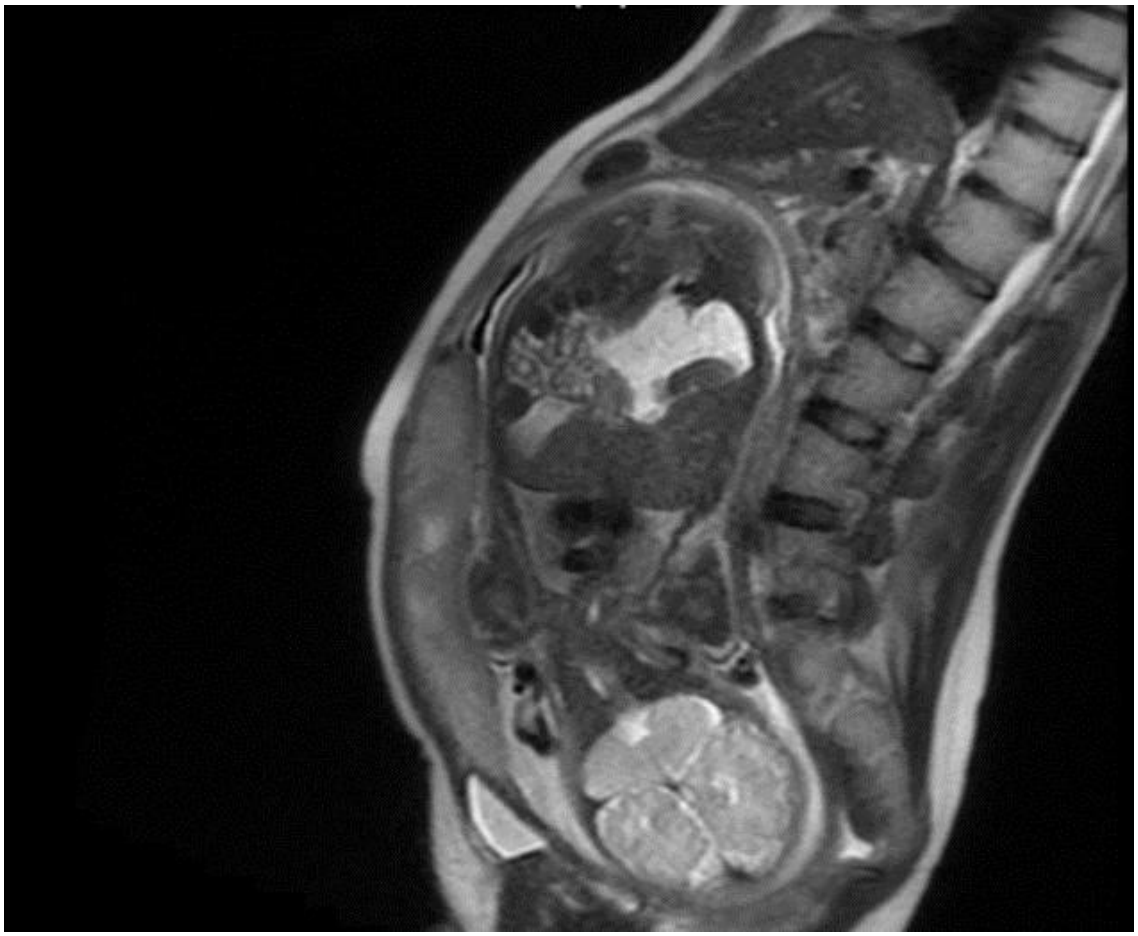


Figure 2 MR sagittal image of the mother showing a large cystic abnormality that is occupying the right half of the foetal abdomen (white arrow).



The case was discussed at the multidisciplinary team meeting and together with the parents a plan for an elective caesarean section at 39 weeks of gestation was made. The mother presented at 37^{+2} weeks with spontaneous rupture of membranes and the baby was delivered via an emergency lower segment caesarean section and was transferred to neonatal paediatric intensive unit for close monitoring.

On examination, the baby was found to be pink, haemodynamically stable with no obvious dysmorphic features and soft abdomen. Chest and abdominal X-rays were normal. An ultrasound of the abdomen was performed on day two of life and a multicystic abnormality was noted to be occupying the right half of the abdomen, extending from the liver edge down to the pelvis measuring approximately 7cm by 4cm by 5cm. This further confirmed the antenatal diagnosis of retroperitoneal lymphangioma.

The baby remained well and was deemed fit for discharge on day four with a plan to repeat ultrasound abdomen after six weeks and a repeat MR abdomen and pelvis in 3 months time.

Discussion

Lymphangioma is a benign hamartoma of the lymphatic system, consisting of multiple dilated vessels. These arise due to a defect in

the development of the lymphatic system, which usually develop from the sixth week of gestation.[3] Lymphangiomas can be classified histologically into 3 main types; simple lymphangiomas consisting of lymphatic capillaries, cavernous lymphangiomas which are made up of larger lymphatic vessels with fibrous adventitia, and cystic lymphangiomas which consist of multiple cysts. All the three types may coexist within the same lesion.[3] Approximately 75% of lymphangiomas are located in the head and neck, while the other 25% involve extremities (11%), trunk (11%), abdomen and genitalia (3%), or mediastinum (1%).[4] Abdominal lymphangiomas are reported to occur most commonly in the mesentery of the small bowel, with the retroperitoneum being the second most common site.[4]

Foetal lymphangiomas are frequently associated with karyotypic or other abnormalities like polyhydramnios, skin oedema and hydrops, thus making prenatal diagnosis important.[5] Although spontaneous regression can sometimes occur in a foetal lymphangioma with normal chromosomes, large lymphangiomas require a perinatal multidisciplinary team approach. Here the possible management options such as prenatal aspiration of the cyst, elective caesarean section, or a delivery mode which will avoid foetal damage are dicussed.[6] Accurate prenatal diagnosis permits a planned delivery, adequate intrapartum monitoring and immediate postnatal resuscitation which could lead to an improved prognosis.

On ultrasonography and MR imaging, cystic lymphangiomas appear as sharply defined, unilocular or multilocular cystic lesions, with thick or thin walled septa. On ultrasound, the fluid may be anechoic, and there may be variable internal echoes or even fluid filled levels, due to bleeding and fibrin deposition.[7] In our case, both antenatal sonography and MR imaging accurately diagnosed the retroperitoneal lymphangioma. The accuracy, safety, immediate availability and its low cost had made ultrasonography the imaging modality of choice in the antenatal assessment for foetal anomalies.[8] However, with the advances in MR imaging, this modality has become a useful tool for the prenatal diagnosis of complex foetal anomalies. In addition, MR imaging has the additional benefit of providing more comprehensive images, accurately delineates the exact extent of the lesion and it is not operator-dependent.[5] MR imaging can also provide improved prenatal parental counselling and postnatal therapeutic planning.[9]

The treatment of choice of retroperitoneal lymphangiomas is complete surgical excision. The long-term prognosis is excellent if complete excision has been achieved. If not, the child needs to be followed up with serial scans to exclude recurrence of the lesion. Another alternative treatment is image-guided percutaneous catheter drainage of lymphangioma followed by sclerotherapy.[10] Successful outcomes have been reported following the use of intralesional bleomycin, sclerotherapy with OK-432, or percutaneous embolization with Ethibloc.[2,11,12]

In conclusion, foetal retroperitoneal lymphangiomas are very rare benign tumours which can be accurately diagnosed prenatally using a combination of ultrasonography and MR imaging. A multidisciplinary perinatal team approach should be used to manage such cases and the prenatal diagnosis is crucial in the management and prognosis of infants with lymphangiomas.

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