Giant axillary macrocystic lymphatic malformation in a newborn

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DEAR SIR

A one-day-old full-term otherwise normal female baby weighing 3000g presented with a mass on her left hemithorax. Antenatal history was unremarkable. Her vitals were all within normal range. On examination, the baby had a large mass measuring 12x15 cm and extended from the sternum to the anterior axillary line (Fig. 1A). The skin overlying the mass had few areas of hyperpigmentation. On palpation, the mass was soft, fluctuant, non-tender, immobile, and non-pulsatile.

Except for an elevated leucocyte count (22 × 109/L) and raised serum bilirubin (total bilirubin: 121.7 µmol/l, direct bilirubin: 19.5 µmol/L), the rest of her labs were normal. She received phototherapy for two days. Her echocardiogram was normal. Ultrasonography showed multiple anechoic lesions with septations without any significant vascular flow. MRI chest revealed a 9.74x5.99x9.91cm multiloculated cystic mass in the left axillary region with extension to left hemithorax; soft tissue is noted which is high in T2WI with areas of fluid-fluid levels suggestive of hemorrhagic changes (Fig. 1B). Extension of mass to the posterior part of left SCM and also the medial side of the arm is depicted with mass effect on the ipsilateral brachiocephalic vein.

With a diagnosis of lymphatic malformation surgical excision was done on the 10th day of life. At surgery, multiple septated cysts were seen with yellow clear content. Excision of cysts was done with some lymphatic tissue left around the axillary region, a tube drain was inserted, and the skin closed in layers. Postoperative recovery was uneventful whereby the drain was removed on day three and the baby was discharged on day seven. Her follow-up visits at the surgical and pediatric clinics were uneventful with normal upper limb movement and function. Histopathology analysis of the excised mass confirmed macrocystic lymphatic malformation (Fig. 1C). The baby is doing fine on a 6-month follow-up (Fig. 1D).

Lymphatic malformations are rare with an occurrence is 1 in 6,000 live births. [1] Half of these lesions are typically present at birth. [2] They are classified as microcystic, macrocystic, and mixed based on the size of the cyst (cut off 2cm). [3] In the index case, it was a macrocystic lesion. They most commonly occur on the neck (70%) followed by the axilla (20%) and frequently on other sites. [4] Presentation depends on the site, the size, complications, and the association with genetic anomalies like Turner's Syndrome, Trisomy 21, and Edwards and Patau syndromes. [5] Diagnosis is made clinically and confirmed on ultrasonography with Doppler, and MRI. Ultrasound with Doppler shows a multicystic lesion with no blood flow in it as found in the index case.

Management of cystic hygroma can be challenging, especially larger ones around the neck due to their...
close proximity to vital structures. Nonetheless, surgical excision is the management of choice due to its effectiveness. [5] Recurrence can occur when complete excision is not achieved. Alternative methods include sclerotherapy, radiotherapy, cryotherapy, electrocautery, and embolization. [5] We opted for surgical excision as the mass was huge and had compressive symptoms on the surrounding structures in addition to a suspicion of intralesional bleed.

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REFERENCES


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