A giant lymphangioma of the body wall in a child: a heavy companion

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DESCRIPTION

A 16-month-old boy presented with a painless swelling of the right chest wall that had been progressively increasing since birth. There was no fever or any other associated symptom. He had been initially evaluated at another centre and diagnosed as having lymphangioma, for which he had received intralesional sclerotherapy (details not available); as the swelling did not respond he was brought to our institute. On examination, a non-tender, well-defined large mass involving the right side of the chest and abdominal wall was found. The mass, measuring 40×38 cm, extended from the anterior midline of the body to the right paraspinal region posteriorly and from the right axilla to the right iliac crest vertically (figure 1). The ratio of the lesion to the total body length of the child was 0.5 in the vertical dimension. The overlying skin had an area of hyperpigmentation and the mass was soft and fluctuant. There was no swelling elsewhere in the body.

MRI showed a large, well-defined lesion with altered signal intensity involving the soft tissues of the anterior, lateral and posterior chest and abdominal wall on the right side (figure 2). The lesion appeared heterogeneously hyperintense on T2-weighted and isointense with focal hyperintensities on T1-weighted images. There were multiple hypointense intralesional septations of varying thickness present. The lesion was seen to reach up to the ribs of the right lateral chest wall but there was no intrathoracic or intra-abdominal extension. Radiological diagnosis of slow-flow vascular malformation was performed.

The boy underwent injection sclerotherapy using a combination of 3% sodium tetradecyl sulfate and oxytetracycline. At a 3-month follow-up it was decided to excise the lesion as there was no significant reduction in the size of the mass. He was placed in a left lateral position and a transverse elliptical incision was made at the middle of the mass extending from the lateral border of the right rectus muscle anterior to the lateral border of the latissimus dorsi posterior. The skin flaps were raised and a near complete excision of the lesion was performed. The mass had multiple cysts containing yellowish brown fluid. A thin superficial layer of lesion was left on the skin flaps to protect skin viability. The closure of the subcutaneous tissue and skin was performed over two suction drains. The patient was discharged uneventfully after the operation.

The histopathology report revealed a cystic lymphangioma. On out-patient follow-up on the eighth postoperative day, fluid collection was found to have developed under the skin flaps. This was managed by repeated aspirations and use of injection bleomycin as local sclerotherapy. The child

remained well at the last follow-up 4 months postoperatively.

Lymphatic malformations (LMs) consist of fluid-filled cysts derived from lymphatic vessels, which fail to develop connection with the draining venous system. They are considered to be a slow flow vascular malformation and classified as microcystic, macrocystic or combined macrocystic and microcystic lesions.¹ The term 'lymphangioma' is frequently used for microcystic LMs. The cervicofacial and axillary region are common sites for LMs.²⁻³ Body wall lymphangiomas are very rare.⁴⁻⁵ The differential diagnosis of such lesions in the paediatric population at this site includes vascular malformations, lipomas, desmoid tumour and malignant soft tissue tumours.⁶ The LMs are usually evident at birth and have progressive enlargement with or without episodic enlargement associated with systemic or localised infection. Clinically, they may present as well-defined localised, ballotable masses or may have extensive tissue involvement. Ultrasound is a useful tool to help initially characterise such mass lesions, however, contrast-enhanced MRI is the most important diagnostic tool as it will show the extent of the lesion and differentiate it from non-vascular and other vascular lesions.¹ Early diagnosis and complete surgical resection is of utmost importance to avoid morbidity and recurrence of such lesions. Injection sclerotherapy is useful in managing LMs as sole therapy and also as an adjunct to surgical management.²⁻³⁻⁷

Figure 1 A large right-sided chest and abdominal wall lymphangioma in a child.

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Figure 2 The MRI scan (coronal (A) and transverse (B) section) of the child with right-sided body wall lymphangioma.

Learning points

- ▶ Lymphangioma of the body wall is a very rare condition that can become extremely enlarged if not managed early.
- ▶ Early complete resection should be undertaken to avoid massive growth and future recurrence.
- ▶ Injection sclerotherapy after surgery is helpful to prevent collection of fluid as well as to prevent recurrence of residual lesions.

Competing interests None.

Patient consent Obtained.

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