Rare disease

A frog in the throat? An expansile mass presenting in the neck with dysphonia

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Summary

A neck mass, which rapidly increases in size over several weeks, is concerning for all involved. When accompanied by other symptoms suggesting sinister underlying pathology, efficient management and rapid diagnosis are vital. The causes may include primary or metastatic carcinoma, or lymphoreticular malignancy. Other non-sinister pathology may account for the swelling such as reactive lymph nodes or benign neoplasms. A benign neoplasm of the lymphatic system known as a lymphangioma may rarely be the cause of a mass in the neck in adults. More commonly found in infants, with approximately 40% found at birth, they are seldom encountered in adults. In children, they have the propensity to infiltrate into and around muscles and neurovascular structures, occasionally leading to difficult surgical excision. The authors present an unusual case of an adult patient with an expansile mass in the supraclavicular fossa of insidious onset. It was associated with dysphagia and hoarseness of voice.

BACKGROUND

We present an unusual presentation of a cervical lymphangioma presenting in an adult with no other preceding history. While common in infants, often as a congenital abnormality, they are uncommon in adults. The rapid expansion of the mass prior to presentation warranted referral under the 2-week wait criteria. There are several case reports of lymphangiomas in adults, often with preceding trauma, but no such history was evident in this case. This is therefore an unusual presentation of a rare disease, not usually seen in this demographic.

CASE PRESENTATION

A middle-aged patient presented with a swelling in the neck, which had been rapidly increasing in size for several weeks. The patient had associated hoarseness of voice, dysphagia with solid food and difficulty breathing. They also noted that on coughing, the swelling appeared to increase in size.

On examination, there was a 5 cm diameter mass in the left supraclavicular fossa. There was no associated erythematous change to the overlying skin, or tenderness on palpation. There were distended superficial neck veins on the left, and chest wall movements on the left were decreased. There was tracheal shift to the right. Bronchial breath sounds were heard over the swelling, and on coughing, the swelling visibly protruded from the supraclavicular fossa. There were no neurovascular deficits to note on examination. A full assessment of the aero-digestive tract was unremarkable.

INVESTIGATIONS

An urgent out-patient CT scan was obtained, which demonstrated a large mass in the left supraclavicular fossa. This was homogenous in nature, and likely to be a lipoma or

fluid-filled cyst. (figure 1) The scan was reviewed at clinic, and aspiration performed. The fluid aspirated composed of small, monomorphic lymphocytes suggesting that the fluid was lymph. The initial report therefore suggested a lymphangioma.

Histological examination showed a convoluted cyst wall lined by endothelium. The lumen was filled with fluid rich in lymphocytes. Significant inflammation or evidence of malignancy was not seen. The histological conclusion was that the lesion was indeed a lymphangioma.

DIFFERENTIAL DIAGNOSIS

The main differential diagnoses included a neoplastic lesion of the lymphoreticular system, which was either benign or malignant, or a pharyngeal perforation. Other diagnoses considered included lipoma, lymphoma, and neoplasms of the lung or pleura.

TREATMENT

The mass was surgically excised, and removed as one large, well circumscribed, fluid filled cyst measuring 50 × 50 × 30 mm. There was no infiltration of the mass into the surrounding structures.

OUTCOME AND FOLLOW-UP

The patient was reviewed at clinic and had made an uneventful recovery. There was no recurrence after 12 months, and was therefore discharged.

DISCUSSION

Lymphangiomas are benign congenital anomalies, typically seen in children. They are rarely seen in adults. They are soft, compressible and slow-growing masses. The term is often used interchangeably with 'cystic hygroma',

Figure 1 CT of thorax showing left-sided lymphangioma with compression of local neurovascular structures. There is some displacement of the larynx away from the midline.

as both neoplasms arise from the lymphatic system, differing only in architecture. Their propensity to infiltrate into and around muscles and neurovascular structures, may lead to difficulty in surgical excision, particularly in children.^{1–8}

Originally thought to be a vascular anomaly in those presenting in infancy, their spontaneous occurrence in adults has given rise to the theory that lymphangiomas are a neoplastic phenomenon.² There is some debate as to the aetiology of lymphangiomas, although several case reports indicate an association with traumatic injury. It is important to note that they may also present in several genetic disorders, including Turner's syndrome and Trisomies 13, 18 and 21. The incidence of cervical lymphangioma is approximately 1 in 500 in both children and adults, but in adults alone the incidence may be as low as 1 in 50 000.^{1 4–6}

The most common site for lymphangiomas are in the cervico-facial region, accounting for two-thirds of lesions, the majority of which appear in the head and neck area, particularly in the posterior triangle. Other common sites include the axilla, chest wall, shoulder, mediastinum, abdominal wall and thigh. The remainder occur throughout the body. Presenting complaints may include cosmetic symptoms and symptoms of mass effect on neurovascular structures.⁷ Clearly, the symptoms will vary depending on the size and location of the lesion. In this case, both cosmetic symptoms and those of mass effect were demonstrated. The impact on local structures was evident on CT scanning.

Lesions presenting in the head and neck require a systematic approach to provide rational differential diagnoses, and therefore an appropriate management plan. In this instance, there were many diagnoses in consideration, including lymphoma, lipoma, lymphangioma, metastatic aerodigestive tract malignancy, and other neoplasms of the neck, lung and pleura. Presentation of a lymphangioma

with dysphagia and hoarseness of voice is unusual. Imaging and biopsy were therefore of paramount importance in ruling out a more sinister pathology.^{1 4 5} Our management aimed to establish the extent of the lesion in order to guide further medical or surgical intervention. As it transpired, there was no evidence of relation to the underlying pleural structures in our case, and the apparent expansile nature of the presentation was simply a result of the position of the cyst, lying directly on the apical pleura.

There is much debate as to the management of lymphangioma. Some centres suggest the use of the sclerosant OK-432 once the diagnosis is confirmed histologically, while others prefer a surgical approach. The surgical approach is more commonly utilised in the UK. Alternatives include a conservative approach, observing for spontaneous regression, aspiration or radiotherapy, with varying degrees of success.^{3 6 7 9–10} Neurovascular involvement or investment is less common in adults, and therefore a surgical approach is more likely to result in complete excision without recurrence.⁸

Identifying the extent of the lesion, and involvement of surrounding structures is of paramount importance in applying management plans. Infiltrative masses are more likely to recur if not excised completely at surgery, whereas well-circumscribed, isolated masses tend not to recur after excision. Infiltrative masses should be imaged prior to being surgically excised, as should recurrent lesions, to ensure eradication of disease. This is reflected in the literature, where surgery is a common treatment.^{1 3 4 6–7} Use of sclerosant therapy appears to be more appropriate in infants but only carries a 76% cure rate.^{3 6 9–11}

The follow-up period in adults is varied, but most institutions report no recurrence at 12 months postsurgical excision. In those with recurrence, this typically occurs within 12 months of treatment. Surgery, however, carries a risk of injury to surrounding structures, including chylothorax, haemothorax, pneumothorax and damage to

neurovascular structures including the marginal mandibular and hypoglossal nerves.^{8–12}

Learning points

- ▶ Although rare in adults, lymphangiomas should be considered when there are expansile cervical masses, or masses presenting with local pressure symptoms.
- ▶ Imaging is essential in choosing the appropriate management of lymphangiomas, to assist planning for complete surgical excision and to prevent recurrence. This allows the surgeon to minimise potential neurovascular injury, and ensure eradication of disease should surgical management become necessary.
- ▶ A follow-up period of 1 year is recommended to ensure there is no recurrence.

Competing interests None.

Patient consent Obtained.

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