Answer

Lymphangioma

Lymphangioma (also known as cystic hygroma) is a benign lymphatic malformation typically found in children aged younger than 2 years.[1] Lymphangiomas are rare lesions with a reported incidence ranging from 2.8:1000 to 1:16 000.[1,2] There are more than 100 adult cases of cystic hygroma reported in the literature.[2,3] While the exact etiology for these lesions is unknown, congenital lymphangiomas are commonly considered to be embryonic malformations of the venous and lymphatic systems.[4] Controversy exists over the origin of adult lymphangiomas; some researchers suggest they can be acquired (eg, through trauma or infection), while others believe they are truly congenital lesions that simply have enlarged. While most lymphangiomas appear in the cervical region, they can occur throughout the body.[5]

As with this patient, lymphangiomas usually present as painless areas of localized swelling.[4] However, with increasing size, they can cause feelings of tightness, compress adjacent structures, bleed, or become infected. Lymphangiomas may grow slowly over years or, in rare instances, develop rapidly over the course of days.[6] While the appearance of adult lymphangiomas may be associated with recent trauma or infection, most cases are idiopathic.[4]

On physical examination, lymphagiomas tend to be fluctuant and mobile. They may transilluminate, but this finding does not occur in most adult cases.[4] Their appearance on examination suggests a differential diagnosis including thyroglossal duct cyst, lipoma, dermoid cyst, laryngocoele, hemangioma, branchial cleft cysts, seromas, metastatic lesions, lymphoma, and ranulas.[3]

Aspirated fluid is usually clear or straw colored.[4,7] Histologic analysis of this fluid typically shows mature lymphocytes with occasional red blood cells and endothelial cells.[7] Imaging is recommended for suspected lymphangiomas to rule out neurovascular involvement or intrathoracic extension. Ultrasound often displays a multilocular cystic lesion. Computed tomographic and magnetic resonance imaging usually show a well-defined cystic mass with fluid attenuation, lobulation, and septation. Based on imaging, lymphangiomas less than 1 cm in diameter can be termed microcysts, while larger lesions are deemed macrocysts.[1]

Surgical excision is the gold standard treatment for adult lymphangiomas.[4] The recurrence rate is 10% to 15%, and recurrence more often occurs in a suprahyoid position or with incomplete excision owing to involvement of adjacent structures such as the carotid artery or esophagus.[4,8] The complication rate is 15% to 30%,with complications including bleeding, infection, lymphorrhea, fistula formation, and neural injury.[4,9]

As some lesions are less amenable to complete surgical excision, other treatment modalities have also been used. Simple aspiration runs a high risk for recurrence. One effective alternative is to sclerose the lesion with OK-432.[1] OK-432 contains lypholized low-virulent group A *Streptococcus pyogenes* and benzylpenicillin, and it generates an inflammatory response, which involutes the cystic space without the induction of scarring.[5] Owing to the induction of an inflammatory response, OK-432 injections can cause fever, pain, localized edema, and rarely cystic enlargement, leading to airway obstruction. These effects usually subside within 5 days. An injection may take 6 weeks to cause a response, and multiple injections may be needed. Only a few reports of successful OK-432 treatment for adult cystic hygromas exist.[1,8] Successful treatment appears to correlate with simple macrocysts between 1 and 5 cm in size located below the mylohyoid.[1] Other sclerosing agents such as alcohol, steroids, bleomycin sulfate, interferon, doxycycline, and cyclophosphamide have been injected into cystic hygromas with varying results.[8] Owing to their less satisfactory recurrence rate, localized tissue damage, or systemic adverse effect profiles, these other sclerosants are rarely used.

This report documents the case of an adult with a very large lymphangioma of idiopathic origin. Despite its size, it was successfully managed with surgery with a satisfactory result. While there is some enthusiasm for the use of OK-432 as a first-line treatment for cystic hygroma (it is currently being studied for Food and Drug Administration approval), for such a large lesion in an easily accessible surgical area, excision would seem the prudent choice.

Accepted for Publication: July 6, 2011.
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Author Contributions: *Study concept and design:* Steigelman. *Acquisition of data:* Steigelman. *Analysis and interpretation of data:* Chauviere. *Drafting of the manuscript:* Chauviere. *Critical revision of the manuscript for important intellectual content:* Steigelman. *Study supervision:* Steigelman.
Financial Disclosure: None reported.

REFERENCES

1. Poldervaart MT, Breugem CC, Speleman L, Pasmans S. Treatment of lymphatic malformations with OK-432 (Picibanil): review of the literature. *J Craniofac Surg.* 2009;20(4):1159-1162.
2. Karkos PD, Spencer MG, Lee M, Hamid BN. Cervical cystic hygroma/lymphangioma: an acquired idiopathic late presentation. *J Laryngol Otol.* 2005;119(7):561-563.
3. Avitia S, Osborne RF. Cystic hygroma exacerbated by pregnancy. *Ear Nose Throat J.* 2005;84(2):78-79.
4. Schefter RP, Olsen KD, Gaffey TA. Cervical lymphangioma in the adult. *Otolaryngol Head Neck Surg.* 1985;93(1):65-69.
5. Cheng LH, Wells FC. A multidisciplinary approach to recurrent cervicothoracic cystic hygroma in an adult. *Br J Oral Maxillofac Surg.* 2004;42(1):66-68.
6. Sherman BE, Kendall K. A unique case of the rapid onset of a large cystic hygroma in the adult. *Am J Otolaryngol.* 2001;22(3):206-210.
7. Henke AC, Cooley ML, Hughes JH, Timmerman TG. Fine-needle aspiration cytology of lymphangioma of the parotid gland in an adult. *Diagn Cytopathol.* 2001;24(2):126-128.
8. Woolley SL, Smith DR, Quine S. Adult cystic hygroma: successful use of OK-432 (Picibanil). *J Laryngol Otol.* 2008;122(11):1260-1264.
9. Charabi B, Bretlau P, Bille M, Holmelund M. Cystic hygroma of the head and neck: a long-term follow-up of 44 cases. *Acta Otolaryngol Suppl.* 2000;543:248-250.