



Contents lists available at ScienceDirect

## International Journal of Surgery Case Reports

journal homepage: [www.casereports.com](http://www.casereports.com)

## Surgical management of adult-onset cystic hygroma in the axilla

Francesca McCaffrey<sup>a,\*</sup>, Joseph Taddeo<sup>b</sup><sup>a</sup> Togus VA Medical Center, Augusta, ME, United States<sup>b</sup> Central Maine Surgical Associates, 12 High Street Suite 401, Lewiston, ME 04240, United States

## ARTICLE INFO

## Article history:

Received 16 October 2014

Accepted 2 November 2014

Available online 11 November 2014

## Keywords:

Cystic hygroma

Lymphangioma

Lymphatic malformation

Axillary mass

## ABSTRACT

**INTRODUCTION:** Lymphatic malformations are commonly recognized as relatively benign congenital masses affecting infants and children in the perinatal period. In children, these masses are most commonly found in the neck, and are occasionally seen in other areas of the body.

**PRESENTATION OF CASE:** A 58-year-old man presented with an acute axillary swelling measuring approximately 20 cm in length, 12 cm in AP width, and 7 cm in depth. Biopsy and cytology analysis demonstrated this mass to be a cystic hygroma of adult-onset.

**DISCUSSION:** Given its multi-loculated nature and size, it was surgically excised and one year later the patient is without evidence of recurrence.

**CONCLUSION:** As the incidence of adult-onset cystic hygroma is rare, the nature and reporting of their management is limited. This case report contributes to the body of literature which serves to elucidate the optimal management of this perinatal condition in adults.

© 2015 Published by Elsevier Ltd. on behalf of Surgical Associates Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/3.0/>).

## 1. Introduction

A cystic hygroma is a malformation of the lymphatic system most often observed in infants and children.<sup>1–3</sup> Case reports of cystic hygroma in the adult are exceedingly rare and a recent report has estimated that fewer than 150 cases can be found in English-language scientific literature.<sup>1</sup> Also referred to as a lymphangiomas or lymphatic malformations,<sup>1–3</sup> cystic hygromas are most often found in the cervicofacial region (75%) and are less frequently seen in the axilla (20%) or elsewhere.<sup>4</sup> Previous reports have discussed the management of cystic hygromas in the adult, but these reviews are limited to presentations in the neck.<sup>5,6</sup> A review of the literature has identified rare case reports of adult-onset cystic hygroma of the axilla.<sup>7–14</sup>

The exact nature of cystic hygroma formation is still debated. Once considered to be neoplastic in nature,<sup>3</sup> these benign masses are now better understood. Approximately one in 2000–4000 lives births is complicated by a cystic hygroma<sup>16</sup> and 90% are diagnosed before 2 years of age.<sup>2,3</sup> Some have suggested the role of trauma or viral infection in their development,<sup>2,15</sup> while others discuss a more congenital etiology involving miscommunication between lymphatic and venous pathways, aberrant lymphatic growths, and tissue sequestration during development. The role of chromosomal abnormalities has been documented, most often involving Turner's

syndrome, trisomies 13, 18, and 21, and Noonan syndrome.<sup>2</sup> A review of the literature did not reveal information about the prevalence of chromosomal abnormalities in adult-onset cases, however.

Several staging and classification mechanisms have been proposed and adopted which allow for better diagnosis and management. Smith et al. described cystic hygromas as either microcystic, macrocystic, or mixed, with microcystic containing cysts less than 2 cm in diameter. This allowed for a more accurate prediction of how the mass would respond to sclerotherapy treatment.<sup>6,17</sup> Mulliken<sup>18</sup> has described cystic hygromas in terms of histological appearance as either capillary lymphangiomas, cavernous lymphangiomas, or cystic hygromas. De Serres<sup>19</sup> proposed a more practical classification system based on location, which allowed for a better estimation of prognosis and surgical complication rate, but applies only to masses in the cervicofacial region. Diagnosis is often aided by the use of fine needle aspiration for cytology, tissue histology, and ultrasound, MRI, or CT for definition of the mass. Loculations or cysts can often be appreciated and affected structures can be identified.<sup>1</sup>

## 2. Case report

A 58-year-old, otherwise healthy man was referred to general surgery for a four month history of a sudden onset, non-tender, right flank and axillary swelling. He reported experiencing no trauma or surgery to that area. Comorbidities included paranoid schizophrenia well-managed with thiothixene, quetiapine, valproate, and benztropine; arthritis controlled with leflunamide, etodolac, and hydroxychloroquine; and hypertension managed

\* Corresponding author at: University of New England, 11 Hills Beach Road, Biddeford, ME 04005, United States. Tel.: +1 518 932 1015.

E-mail addresses: [fmerchant@une.edu](mailto:fmerchant@une.edu) (F. McCaffrey), [joemoves@yahoo.com](mailto:joemoves@yahoo.com) (J. Taddeo).

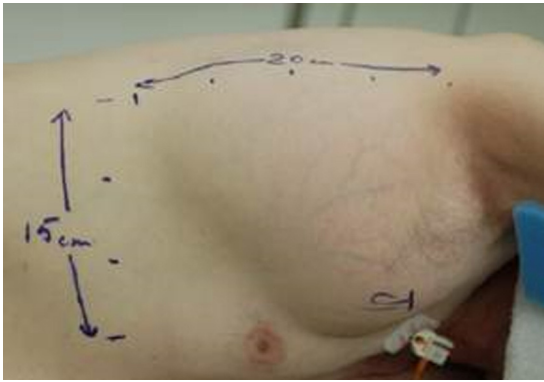


Fig. 1. Mass as demonstrated on physical examination.

with lisinopril. His only prior operative procedure was an open appendectomy as a child. He had no history of substance abuse and his available family history was unremarkable for congenital masses or chromosomal abnormalities. On physical examination, he was well-nourished and well-developed, with normal temperature, vital signs and cardiopulmonary exam. He was found to have a 20 cm × 12 cm × 7 cm, fluctuant, non-tender, collection over the right upper flank extending superiorly into the axilla and anteriorly onto the lateral aspect of the pectoralis major (Fig. 1). Magnetic resonance imaging demonstrated a large multi-septated non-enhancing cystic mass associated with the right axilla and lateral chest wall (Fig. 2). He was scheduled for a laparoscopic aspiration and tissue biopsy under general anesthesia and, after confirmation of the diagnosis, underwent surgical excision of the tumor. At one year post-op, the patient is without any evidence of recurrence or complaints.

Initial biopsy and cytological analysis for diagnostic confirmation was performed under general anesthesia with the patient in the left lateral decubitus position. A full-thickness sample of the capsule wall was removed for analysis. Laparoscopic guidance was utilized to sample various internal parts of the lesion through a minimal incision and was helpful in identifying and lysing additional loculations (Fig. 3). A total of 1450 cc of clear, straw-colored fluid was recovered and sent for gram stain, culture and cytology. He tolerated the procedure without pain and although completely drained, the lesion reaccumulated a week later.

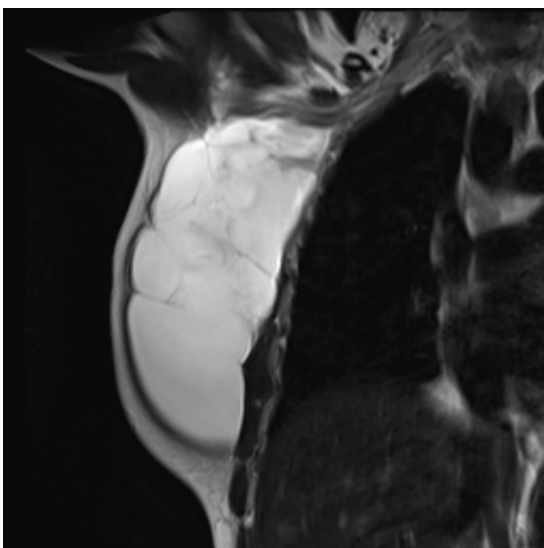


Fig. 2. Magnetic resonance imaging demonstrating a large multi-septated non-enhancing cystic mass.

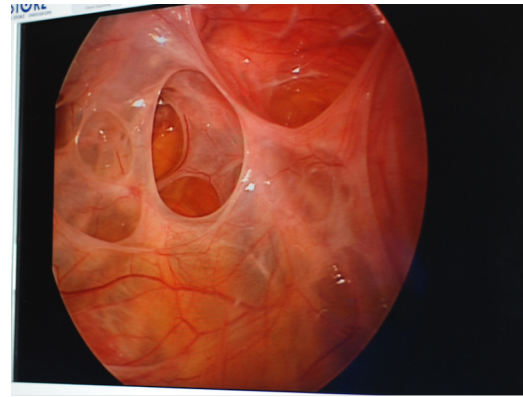


Fig. 3. Laparoscopic view of internal septations during initial aspiration and tissue biopsy.

Four weeks after the initial biopsy and with confirmation of the diagnosis, a full excision of the mass was performed. The patient was positioned in the same fashion and the procedure began with an elliptical incision which encompassed the two prior incisions and provided an anchor for removal of the cystic hygroma. It was dissected away from the overlying latissimus dorsi muscle posterolaterally and serratus anterior muscle anteriorly. Where the capsule wall was thin and friable, suture ligation was necessary to prevent spillage of contents and maintain integrity of the structure to ensure its complete removal. As the dissection continued cephalad, the medial pectoral, long thoracic, and thoracodorsal nerves were identified and spared. Where the cyst became embedded in the axilla, a gentle blunt dissection was sufficient to remove the remaining mass along with a few incorporated lymph nodes (Fig. 4). The skin was closed in two layers of absorbable sutures and drained with two 10 mm Jackson-Pratt drains. The patient was observed for four days due to psychosocial circumstances, and the drains were removed on the day of discharge. The patient agreed to annual follow-up.

Cytologic analysis of the initial fine needle aspiration revealed lymphocytes and scarce red blood cells consistent with a diagnosis of a cystic hygroma. The full thickness biopsy of the mass capsule revealed simple cuboidal epithelium, which stained positive for D2-40 and CD31, as expected.<sup>8</sup> Tissue samples from inside the mass returned the same report. The specimen provided after excision measured 17 cm in length by 10 cm in depth with an average thickness of 3 cm. The mass was observed to have many loculations consistent with a cystic hygroma of both micro- and macrocystic anatomy.

Two weeks after surgical excision, the patient presented for physical examination which was remarkable only for minimal



Fig. 4. Cystic hygroma, ex vivo, measuring 17 cm × 10 cm × 3 cm.

superficial numbness at the incision site. Motor strength was preserved in all muscle groups.

### 3. Discussion

Differential diagnosis for a cystic hygroma includes soft tissue sarcoma, abscess, synovial cyst, and hematoma.<sup>18</sup> Although it was strongly suspected on initial imaging to be a cystic hygroma, given the extreme rarity of this condition in adults, we chose to ensure that this was not an unusual presentation of a neoplasm by performing a biopsy and aspiration.

Treatment of a cystic hygroma has historically required surgical excision,<sup>1–3,5</sup> which some believe to be preferred in adults as lesions are better circumscribed.<sup>3,15</sup> In one case report, surgical excision was felt to be particularly uncomplicated with thin encapsulation of the mass, weak adhesions to surrounding tissues, with minimal neurovascular sacrifice.<sup>1</sup> Further, the potential for spontaneous bacterial infection heightens the risk of delaying therapy in hopes of achieving the spontaneous regression sometimes seen in children.<sup>3</sup> A reasonable concern about surgical therapy is the possibility of recurrence to compound the risks of the procedure. Indeed, some authors have discussed the role of intraoperative cyst rupture making the boundaries of the mass less clear.<sup>20</sup> However, one study found that in patients known to have partial resection of their mass, only one in nine had recurrence.<sup>21</sup> Several case reports have demonstrated the inefficacy of simple aspiration or antibiotics.<sup>1,9,13</sup> Long term follow-up has been suggested as an important element of management, as recurrence has occurred as late as 6 years after removal.<sup>15</sup>

Some authors have emphasized the role of sclerotherapy in cystic hygroma management,<sup>2</sup> using agents such as OK-432, bleomycin, doxycycline, acetic acid, alcohol, and hypertonic saline.<sup>5</sup> This therapy was not pursued for several reasons given the proximity of the mass to vital neurovascular structures and mixed cystic anatomy. First, the success of sclerotherapy has occasionally been measured in terms of sufficient mass reduction without complete resolution.<sup>1</sup> Further, it has been recognized that sclerotherapy may not be effective against multiloculated masses, or those of mixed or microcystic anatomy.<sup>1,2,5</sup> Sclerotherapy may also induce a localized immune response which causes a temporary but dramatic increase in the size of the mass. Perkins et al. has also recognized the potential for shock-like reaction in the setting of penicillin allergy and OK-432 use.<sup>5</sup>

### 4. Conclusion

Continued reporting of adult-onset cystic hygroma is of particular importance, as the nature and management of these rare masses are elucidated. The role of chromosomal abnormalities in cases presenting in the adult is not yet understood and may be of interest as such masses continue to be reported. This case contributes to the body of evidence supporting the role of cystic hygroma in a differential diagnosis for masses in the adult, especially in the acute setting. Further, the future management of cystic hygromas in the axilla, with proximity to important neurovascular structures, is better informed with our addition to examples of uncomplicated resection.

### Conflict of interest

The authors have no conflicts of interest to report.

### Open Access

This article is published Open Access at [sciendo.com](http://sciendo.com). It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.

### Funding

None.

### Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

### Author contributions

Joseph Taddeo contributed to the manuscript editing and Francesca McCaffrey contributed to the manuscript writing.

### Acknowledgments

The authors acknowledge Dr. Robert Lenox for his guidance in the submission of this manuscript.

### References

- Gow L, Gulati R, Khan A, Mihameed F. Adult-onset cystic hygroma: a case report and review of management. *Grand Rounds* 2011;**11**(1):5–11.
- Bloom DC, Perkins JA, Manning SC. Management of lymphatic malformations. *Curr Opin Otolaryngol Head Neck Surg* 2004;**12**:500–4. PMID: 15548907.
- Naidu SI, McCalla MR. Lymphatic malformations of the head and neck in adults: a case report and review of the literature. *Ann Otol Rhinol Laryngol* 2004;**113**:218–22. PMID: 15053205.
- Sarin YK. Cystic hygroma. *Indian Pediatr* 2000;**37**:1139–40.
- Perkins JA, Manning SC, Tempero RM, Cunningham MJ, Edmonds JL, Hoffer FA, et al. Lymphatic malformations: review of current treatment. *Otolaryngol Head Neck Surg* 2010;**142**:795–803.
- Adams MT, Saltzman B, Perkins JA. Head and neck lymphatic malformation treatment: a systematic review. *Otolaryngol Head Neck Surg* 2012;**147**(4):627–39.
- Philippakis GE, Manoloudakis N, Marinakis A. A rare case of a giant cavernous lymphangioma of the chest wall and axilla in an adult patient. *Int J Surg Case Rep* 2013;**4**(2):164–6.
- Krainick-strobel U, Krämer B, Walz-mattmüller R, Kaiserling E, Rohm C, Bergmann A, et al. Massive cavernous lymphangioma of the breast and thoracic wall: case report and literature review. *Lymphology* 2006;**39**(3):147–51.
- Michail O, Michail P, Kyriaki D, Kolindou A, Klonaris C, Griniatsos J. Rapid development of an axillary mass in an adult: a case of cystic hygroma. *South Med J* 2007;**100**:845–9.
- Stoss S, Kalbermatten DF, Robertson A, Bruder E, Rasmus M, Gambazzi F, et al. Large cystic tumour at the chest wall mimicking an echinococcosis: a case report. *J Plast Reconstr Aesthet Surg* 2008;**61**:13–6.
- Nguyen K, Karsif K, Lee S, Chorny K, Chen M. Lymphangioma in an elderly patient: an unusual cause of axillary mass. *Breast* 2011;**17**:416–26.
- Gebrim LH, de Lima GR, Tanaka CI. Axillary cystic lymphangioma in pregnancy. *Int J Gynaecol Obstet* 1995;**48**:327–8.
- Katharina C, Quack L, Abdul-Raham J, Elisabeth G, Judit P, Ernst B. Axillary cavernous lymphangioma in pregnancy and puerperium. *Gynecol Obstet Invest* 2005;**60**:108–11.
- Smith RC, Sherck HH, Kollmer C, Javitt MC. Cystic lymphangioma in the adult: an unusual axillary mass. *Magn Reson Imaging* 1989;**7**:561–3.
- Schefter RP, Olsen KD, Gaffey TA. Cervical lymphangioma in the adult. *Otolaryngol Head Neck Surg* 1985;**93**(1):65–9.
- Kennedy TL, Whitaker M, Pellitteri P, Wood WE. Cystic hygroma/lymphangioma: a rational approach to management. *Laryngoscope* 2001;**111**(11, Pt. 1):1929–37.
- Smith RJ, Burke DK, Sato Y, Poust RI, Kimura K, Bauman NM. OK-432 therapy for lymphangiomas. *Arch Otolaryngol Head Neck Surg* 1996;**122**(11):1195–9.
- Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: a classification based on endothelial characteristics. *Plast Reconstr Surg* 1982;**69**(3):412–22.
- De serres LM, Sie KC, Richardson MA. Lymphatic malformations of the head and neck. A proposal for staging. *Arch Otolaryngol Head Neck Surg* 1995;**121**(5):577–82.
- 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):206–10.
- Riechelmann H, Muehlhays G, Keck T, Mattfeldt T, Rettinger G. Total, subtotal, and partial surgical removal of cervicofacial lymphangiomas. *Arch Otolaryngol Head Neck Surg* 1999;**125**(6):643–8.