Case Report

Access this article online
Website: www.afrjpaedsurg.org
DOI: 10.4103/0189-6725.78937
Quick Response Code:

Cystic hygroma of unusual sites: Report of two cases

Bilal Mirza, Lubna Ijaz, Shahid Iqbal, Ghulam Mustafa, Muhammad Saleem, Afzal Sheikh

ABSTRACT

Cystic hygroma is the cystic variety of lymphangiomas. Its common site is cervico-facial region, followed by axilla, superior mediastinum, mesentery and retroperitoneal region. Its occurrence in upper extremity is rarely reported, whereas its location at sternum has not been reported before. We report two cases of cystic hygroma occurring at unusual sites.

Key words: Bleomycin, cystic hygroma, lymphangioma, unusual site

INTRODUCTION

Lymphangioma is the congenital malformation of lymphatic system, resulting from failure of development of communications between lymphatics of various regions.[1]

Eighty percent of cystic hygromas reside in cervico-facial region. Other sites are axilla, superior mediastinum, mesentery, retroperitoneum, pelvis, and lower limbs. Upper extremity is a rare site where cystic hygromas can be located.[2,3] Literature regarding this location of cystic hygroma is scarce. Only a few case reports have described its occurrence in the arm and forearm.[4,5] The occurrence of cystic hygroma at sternal site is very rare, and to the best of our knowledge, has not been reported before.

CASE REPORTS

Case 1

A 2-month-old male infant presented to the OPD of our institution with complaints of swelling at right arm and forearm since birth. There were no other complaints. On examination, the swelling was extending from upper one-third of arm to lower two-thirds of forearm on anterior, lateral and posterior aspects [Figures 1 and 2]. The swelling was non-tender and there were no signs of acute inflammation present. The swelling was palpated to have multiple cystic areas in it which were easily



Figure 1: (Case 1) Cystic mass on the antero-lateral aspect of arm and forearm. In this view, two major cysts are evident



Figure 2: (Case 1) Another view of the same patient showing posterior extension of the cystic hygroma

Department of Pediatric Surgery, The Children's Hospital & The Institute of Child Health, Lahore, Pakistan

Address for correspondence:
Dr. M. Bilal Mirza, Department of Pediatric Surgery, The Children's Hospital & The Institute of Child Health, Lahore, Pakistan.
E-mail: blmirza@yahoo.com

compressible and transluminant. The patient already had magnetic resonance imaging (MRI) done, as per the advice of some other institution. MRI scan showed huge cystic areas involving right arm, predominantly antero-lateral aspect, with multiple septations showing TW1/TW2 prolongation, suggestive of cystic hygroma. Post-contrast images revealed enhancement through the septations. There was no bony involvement found [Figure 3].

Our diagnosis after clinical examination was cystic hygroma supported by MRI. Patient was put on sclerosant therapy that showed significant reduction in size of lesion, after three sessions of sclerotherapy with bleomycin at a dose 0.5 mg/kg [Figure 4].

Case 2

A 5-year-old child was presented to OPD of our institution with complaints of swelling in front of sternum since birth. Initially, the swelling was static in size but started increasing in size for a few months. On clinical examination, an isolated cystic swelling of about 7 cm × 7 cm size, in front of sternum, not attached to the underlying bone, transluminant and without signs of inflammation, was found [Figure 5]. Ultrasound swelling revealed a 6.5 × 6 cm cystic lesion with internal septations having no intra-thoracic extension. On color Doppler, no flow was identified. Clinical diagnosis of cystic hygroma developed and the patient was advised aspiration and sclerotherapy. On aspiration with wide bore needle, chylous fluid was found, thus confirming our diagnosis [Figure 6]. The patient responded well to the sclerotherapy and only a small fibrotic thickening of the area was left behind.

DISCUSSION

Hygroma is from Greek meaning "moist or watery tumour". Cystic hygroma is the most frequent

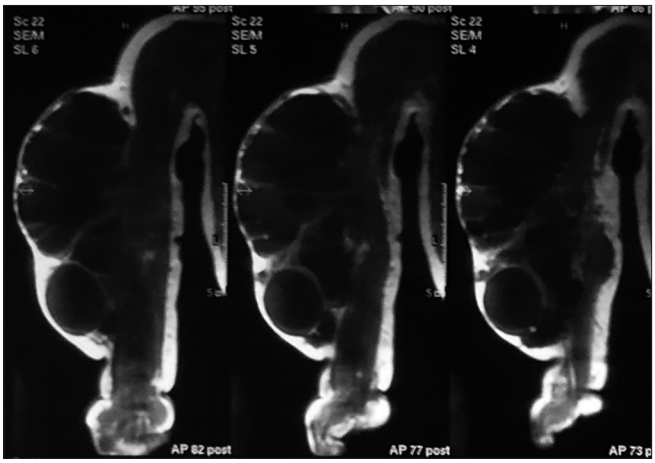


Figure 3: (Case 1) MRI scan of the cystic lesion in arm and forearm. Multiple macrocystic lesions in that location are seen



Figure 4: (Case 1) Marked reduction in size of cystic hygroma can be seen after three sessions of sclerotherapy

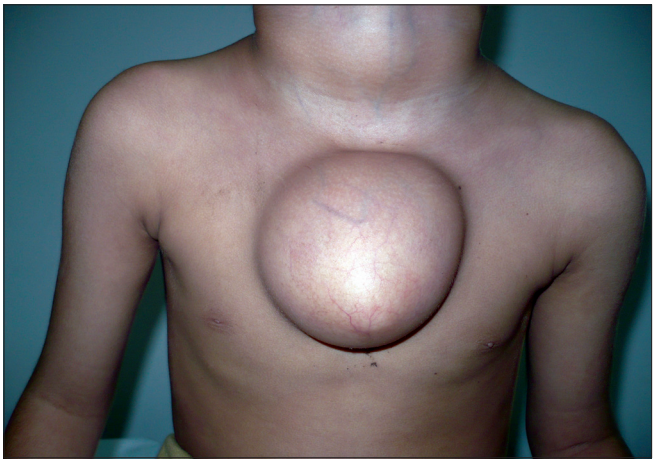


Figure 5: (Case 2) Cystic hygroma in front of sternum in a 5-year-old boy

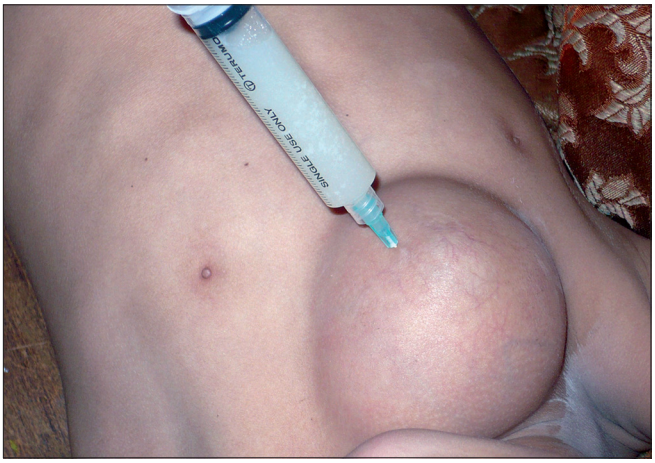


Figure 6: (Case 2) Chyle is the yield while aspirating the lesion

type of lymphangiomas resulting from congenital maldevelopment of lymphatic system. During the course of embryogenesis of lymphatic system, six primary lymphatic sacs are formed; two jugular, two iliac, one retroperitoneal and one dorsal to the retroperitoneal sac, the cisterna chyli. Later on, numerous lymphatic channels from head, neck, limbs, and body wall join the lymphatic sacs. Any maldevelopment or event resulting in failed or insufficient communication between lymphatics of various organs results in the indexed entity. Lymphangiomas may be classified as simple (capillary), cavernous and cystic. Simple lymphangiomas are composed of capillary-sized lymphatic channels with considerable connective tissue stroma. Cavernous lymphangiomas are composed of actively growing, dilated lymphatic channels in a lymphatic stroma. Cystic lymphangiomas or cystic hygromas are single or multiple macrocystic lesions having scarce communication with normal lymphatic channels.[6,7]

Most of the cystic hygromas are evident clinically before the end of infancy; however, their nascent presentation in adults is also reported.[4]

Approximately 80% of all cystic hygromas involve the cervico-facial region. Other sites are oral cavity, axilla, mediastinum, abdominal cavity, retroperitoneum, scrotum and even skeleton. Their location in upper extremity is very rare and confined to a few case reports.[5] However, the location of cystic hygroma at sternum is not documented before.

Macroscopically, cystic hygromas are multiloculated, multilobular or macrocystic masses composed of many individual cysts. Microscopically, the cyst walls consist of a single layer of flattened epithelium.[6] In the first patient, the cystic hygroma of the limb was composed of multiple macrocystic masses, easily delineated by inspection and palpation, whereas in the second patient with cystic hygroma in front of sternum, it was composed of single cystic area with internal septations. The diagnosis of cystic hygroma in the first patient was obvious at clinical examination; however, in the second patient, the lesion masqueraded as a lipoma or dermoid cyst. Doppler ultrasound and chylous aspirate facilitated the diagnosis in that case.

The cysts may contain milky, serous, serosangious or straw-coloured fluid. Cystic hygroma is usually a transluminant lesion; however, it may turn opaque when there is haemorrhage or infection in it.[3,6,8]

Cystic hygromas are benign lesions. However, sometimes complications can arise. The reported complications are haemorrhage inside cysts, infection, spontaneous or traumatic rupture, nerve compression causing pain and paraesthesias, respiratory difficulty, dysphagia, and disfigurement.[1,2]

The differentials of cystic lesions of chest lying in midline or over sternum are dermoid cyst, lipoma, neurofibroma, ectopia cordis, etc.[8] In our case, the swelling was transluminant, non-pulsatile, non-tender, and yielded chyle on aspiration, thus fulfilling the criterion of cystic hygroma in front of sternum.

The management options are surgical excision or sclerosant therapy. The recurrence rates are high even after apparent complete excision of the lesion.[3,6,8,9] Sclerotherapy as a primary treatment modality, for such lesions, is under trial in many centres; many case reports and original studies with good response to the therapy, started appearing in the literature since the last two decades. Most of the studies documented complete resolution of the lesion in about 60–80% of cases. The reported complications of sclerotherapy with bleomycin were non-lethal in these studies and restricted to mild discolouration of the skin at the injection site.[10-13]

We perform both surgical excision and sclerosant therapy for cystic hygroma in our institution. Most of the time, combined surgical excision and sclerosant therapy has proved a boon for the patients having cystic hygroma extended to major vessels or vital structures.

To conclude, upper extremity and sternum as location of cystic hygromas are a rarity and should be considered in the differentials of cystic lesions of the limbs and midline chest wall. Sclerotherapy with bleomycin is a safer and cosmetically acceptable option of management for such lesions.

REFERENCES

1. Manikoth P, Mangalore GP, Megha V. Axillary cystic hygroma. J Postgrad Med 2004;50:215-6.
2. Mirza B, Ijaz L, Saleem M, Sharif M, Sheikh A. Cystic hygroma: An overview. J Cutan Aesthet Surg 2010;3:139-44.
3. Kocher HM, Vijaykumar T, Koti RS, Bapat RD. Lymphangioma of the chest wall. J Postgrad Med 1995;41:89-90.
4. Dhrif AS, El Euch D, Daghfous M, Cherif F, Mokni M, Dhahri AB. Macrocystic lymphatic lymphangioma (cystic lymphangioma) of upper extremity: A case report. Arch Pediatr 2008;15:1416-9.
5. Carpenter CT, Pitcher JD Jr, Davis BJ, Gomez R, Schofield TD, Youngberg RA. Cystic hygroma of arm: A case report and review of literature. Skeletal Radiol 1996;25:201-4.
6. Mansingani S, Desai N, Pancholi A, Prajapati A, Vohra PA, Raniga S. A case of axillary cystic hygroma. Indian J Radiol Imaging 2005;15:517-9.

7. Sadler TW In: Sadler TW, editor. Langman's medical embryology. 9th ed. Philadelphia, Lippincott Williams and wilkins; 2004. p. 269-70.
8. Oak SN, Redkar RG, Kapur VK. Posterior midline cervical fetal cystic hygroma. J Postgrad Med 1992;38:93-6.
9. Arora A, Narula MK, Sonkar P, Chadha R. Cystic hygroma of chest wall. Indian J Radiol Imaging 2003;13:120-1.
10. Ibrahim AH, Kandeel A, Bazeed MF. Successful non surgical management of a huge life threatening cervicomediastinal cystic hygroma case report and review of the literature. J Pediatr Surg Specialt 2009;3:48-50.
11. Okada A, Kubota A, Fukuzawa M, Imura K, Kamata S. Injection of bleomycin as a primary therapy of cystic lymphangioma. J Pediatr Surg 1992;27:440-3.
12. Orford J, Barker A, Thonell S, King P, Murphy J. Bleomycin therapy for cystic hygroma. J Pediatr Surg 1995;30:1282-7.
13. Zhong PQ, Xhi FX, Li R, Xue JL, Shu GY. Long-term results of intratumorous bleomucin-A5 injection for head and neck lymphangioma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1998;86:139-44.

Cite this article as: Mirza B, Ijaz L, Iqbal S, Mustafa G, Saleem M, Sheikh A. Cystic hygroma of unusual sites: Report of two cases. Afr J Paediatr Surg 2011;8:85-8.

Source of Support: Nil, **Conflict of Interest:** None declared.

Staying in touch with the journal

1) Table of Contents (TOC) email alert
Receive an email alert containing the TOC when a new complete issue of the journal is made available online. To register for TOC alerts go to **www.afrjpaedsurg.org/signup.asp.**

2) RSS feeds
Really Simple Syndication (RSS) helps you to get alerts on new publication right on your desktop without going to the journal's website. You need a software (e.g. RSSReader, Feed Demon, FeedReader, My Yahoo!, NewsGator and NewzCrawler) to get advantage of this tool. RSS feeds can also be read through FireFox or Microsoft Outlook 2007. Once any of these small (and mostly free) software is installed, add **www.afrjpaedsurg.org/rssfeed.asp** as one of the feeds.