

Cystic lymphangioma of the breast in an infant successfully managed with intralesional bleomycin: a case report with relevant review of the literature

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Cystic hygromas, also known as lymphangiomas, are unusual congenital malformations of the lymphatic system and commonly involve the head and neck region or axilla. Involvement of other sites such as breasts is very rare. The preferred mode of treatment for lymphangioma of the breast in adults or children is surgery. We report a case of breast lymphangioma in a 3-month-old male child, which was managed successfully by intralesional bleomycin.
Ann Pediatr Surg 13:163–165 © 2017 Annals of Pediatric Surgery.

Annals of Pediatric Surgery 2017, 13:163–165

Keywords: breast, bleomycin, intralesional sclerosing agent, macrocystic lymphangioma

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Received 24 June 2016 accepted 4 September 2016

Introduction

Cystic lymphangioma (CL) is a multiloculated congenital malformation of the lymphatic system, occurring in approximately one in 6000–12 000 births. Almost all cases are diagnosed by the age of 2 years [1]. This term comes from the Greek word 'hygroth' meaning fluid and 'oma' meaning tumor. Cystic hygromas are slow-growing benign tumors, resulting from a developmental anomaly of the lymphatic system. It can affect any site of the body, but is seen more commonly in the head and neck region and the axilla. It is also reported to occur in the mediastinum, retroperitoneum, breasts, and other regions [2]. Treatment of CL occurring at unusual sites by large is complete surgical excision if possible. We report a case of an infant with breast CL, which was managed successfully by intralesional bleomycin.

Case report

A 3-month-old male child weighting 5 kg presented with a lump in the breast (Fig. 1). The lump progressively increased in size and was not painful with no associated nipple discharge. No history of trauma or previous surgery was reported. On palpation, the lump was mobile and nontender and involved almost the entire left breast with no significant axillary lymphadenopathy. His general physical and systemic examinations were unremarkable. Laboratory examinations including complete blood count, serum electrolytes, liver function tests, alkaline phosphatase levels, and thyroid function tests were all within normal limits. Ultrasonography of the breast showed a single large cystic lesion, located in the lateral half of the breast, suggestive of lymphangioma (Fig. 2). Aspiration under ultrasound guidance showed chylous fluid. Considering the cystic nature and the presence of chylous fluid on aspiration, the diagnosis of CL was made. As per our treatment protocol, the patient was advised for intralesional sclerotherapy. At our center, we use aqueous Bleomycin solution as a sclerosing agent, because it is cheap, easily available, effective, and safe. All interventions are performed under general anesthesia/sedation. The cyst was punctured

under USG guidance, the content of the cyst was aspirated completely, bleomycin was injected into the cyst, and care was taken that the total dose should not exceed the recommended dose [0.5 mg/kg (3 mg/ml solution)]. After the procedure, compression of the injection site was performed for 6 h. Strict monitoring of vitals was carried out up to 6 h after the procedure, and the patient was discharged after 24 h. At our center, we review every case after 3 months, and the second session of review is carried out only after 6 months if required. The patient was reviewed after 3 months: clinically no palpable lump was detected (Fig. 3), and ultrasound examination showed a small cystic lesion that was not amenable for a second session. The patient is doing well at the follow-up after 2 years.

Discussion

CL is a congenital malformation that likely results from sequestration of lymphatic tissue that fails to communicate with the rest of the lymphatic system. The origin of lymphangiomas is not clearly understood. Most authors favor the theory that dysplastic lymphatic tissue is sequestered in a target tissue during fetal development [3], whereas abdominal trauma, inflammatory processes, surgery, or radiation therapy may lead to recurrence of such tumors [4]. They can occur anywhere in the body, but 75% involve the posterior neck, 20% the axilla, and 1% the mediastinum, groin, retroperitoneum, and breasts [5]. The breast is a very infrequent site for the occurrence of the cystic subtype of lymphangiomas, with very few cases documented in the literature and is extremely rare in neonates [5].

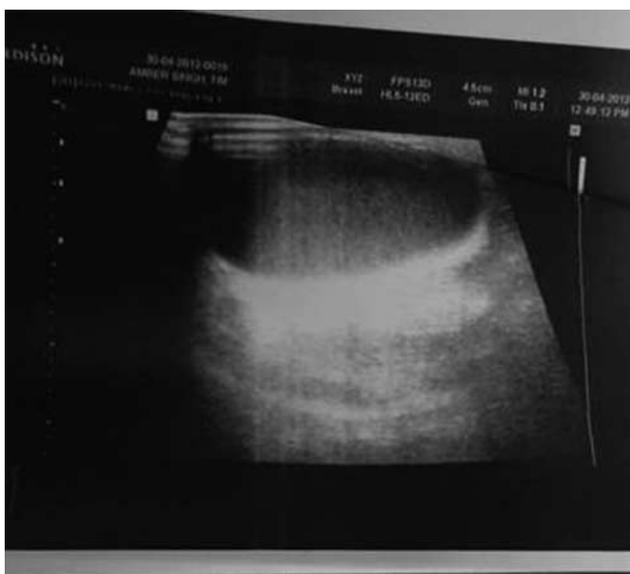
Breast lymphangiomas tend to occur in the upper outer quadrant of the breast [6], whereas in our case it involved the whole breast. This pattern of location may be related to the drainage pattern of the lymphatics in the breast, which is mainly toward the tail and the axillary region [6,7]. CL is mainly a childhood entity under extremely rare instances when the breast is the primary location; most of such cases have been reported in adults [7,8]. Regarding the age distribution of CL, it has

Fig. 1



Cystic lesion in the left breast.

Fig. 2



Ultrasound showing the cystic lesion.

been reported usually in young females ranging from 4 months to 49 years of age [9]. Until now, only five [5,9–12] cases (Table 1) of CL of the breast have been reported in the world literature, of which only one was an infant.

A typical lesion of CL may consist of a single or an aggregation of cysts; usually the larger cysts are located in the periphery, whereas the smaller ones lie deeper and may infiltrate in the neighboring structures [6]. On histology, they are characterized by a single layer of endothelium containing, usually, a clear watery fluid (lymph). CLs are benign with no malignant potential, painless, soft, and fluctuant malformations that may or may not increase on coughing or crying and is brilliantly translucent if the content is clear fluid [6]. Cystic hygromas are benign lesions, but warrant urgent attention because of the complications leading to sudden increase

Fig. 3



Patient after 2 years of follow-up.

in size. Rapid enlargement may occur, usually because of hemorrhage into the cyst, inflammation, or trauma.

The differential diagnoses of CL of the breast include simple cyst, abscess, hematoma, postoperative fluid collection, and hemangioma. Simple cysts tend to occur in both breasts and can often occur in other regions of the same breast and the contents are turbid yellow or green. Hemangiomas generally show thick-walled blood vessels with numerous red blood cells that can be differentiated by Doppler ultrasound [3].

In the head and neck region, clinical signs and symptoms can confirm the diagnosis of CL. However, in atypical places such as breasts, confirmation requires imaging modalities such as Doppler ultrasound/MRI. Ultrasound can usually confirm and differentiate cystic lymphatic malformation with other cystic lesions. On ultrasonography, CLs are seen as cystic multiloculated masses, with linear septa of variable thickness that may contain solid components, although in certain circumstances.

Our review of the literature revealed that CL of the breast is mostly surgically excised. As these lesions are locally aggressive and tend to infiltrate the surrounding tissues, achieving safe surgical margins may at times be difficult. To achieve complete clearance, wide local excision including sacrifice of normal breast tissue can lead to poor, long-term aesthetic and functional outcomes. Keeping all these points in mind and our experience of bleomycin sclerotherapy in the head and neck region, intralesional bleomycin sclerotherapy was attempted after

Table 1 Children with cystic lymphangioma of breast and their management

References	Age (male)	Sites	Management	Follow-up
Kangesu [9]	6 year	Left breast (no details available)	No details available	No details available
Chiba et al. [5]	4 months	Left breast (no details)	Surgical excision	12 months
Singh et al. [12]	6 year	Left breast (upper outer quadrant)	Surgical excision	18 months
Gupta et al. [11]	8 year	Right breast (upper outer quadrant)	Surgical excision	9 months
Ekmez et al. [10]	5 year	Left breast (upper half of breast)	Surgical excision	12 months
This study	3 months	Left breast (whole breast)	Intralesional sclerotherapy	24 months

informed consent in our patient with equivocal results without scar.

We believe that intralesional bleomycin sclerotherapy can be safely used for CL of the breast without long-term complications.

Conflicts of interest

There are no conflicts of interest.

References

- 1 Bill AH Jr, Sumner DS. A unified concept of lymphangioma and cystic hygroma. *Surg Gynecol Obstet* 1965; **120**:79–86.
- 2 Torcasio A, Veneroso S, Amabile MI, Biffoni M, Martino G, Monti M, et al. Cystic hygroma of the breast: a rare lesion. *Tumori* 2006; **92**:347–350.
- 3 Zadvinskis DP, Benson MT, Kerr HH, Mancuso AA, Cacciarelli AA, Madrazo BL, et al. Congenital malformations of the cervicothoracic lymphatic system: embryology and pathogenesis. *Radiographics* 1992; **12**:1175–1189.
- 4 Bossert T, Gummert JF, Mohr FW. Giant cystic lymphangioma of the mediastinum. *Eur J Cardiothorac Surg* 2002; **21**:340.
- 5 Chiba T, Ibrahim M. Cavernous lymphangioma of the breast: case report of an infant. *Nippon Geka Hokan* 1995; **64**:23–26.
- 6 Chung SY, Oh KK, Kim DJ. Mammographic and sonographic findings of a breast cystic lymphangioma. *J Ultrasound Med* 2003; **22**:307–309.
- 7 Ogun GO, Oyetunde O, Akang E. Cavernous lymphangioma of the breast. *World J Surg Oncol* 2007; **5**:69–72.
- 8 Sa EJ, Choi YH. Cystic lymphangioma of the breast. *J Clin Ultrasound* 1999; **27**:351–352.
- 9 Kangesu T. Cystic hygroma of the breast in childhood. *Br J Clin Pract* 1990; **44**:787–788.
- 10 Ekmez F, Pirgon O, Bilgin H, Aydemir G. Cystic hygroma of the breast in a 5 year old boy presenting as a gynecomastia. *Eur Rev Med Pharmacol Sci* 2012; **16** (Suppl 4):55–57.
- 11 Gupta SS, Singh O. Cystic lymphangioma of the breast in an 8-year-old boy: report of a case with a review of the literature. *Surg Today* 2011; **41**:1314–1318.
- 12 Singh O, Singh Gupta S, Upadhyaya VD, Sharma SS, Lahoti BK, Mathur RK. Cystic lymphangioma of the breast in a 6-year-old boy. *J Pediatr Surg* 2009; **44**:2015–2018.