Giant Lymphangioma Breast – A Rare Occurrence: Case Report and Current Review

Abstract

Cystic lymphangiomas are rare benign tumor and are due to congenital malformations of lymphatic vessels. Majority occur either at birth or up to 2 years age. These are slow-growing lesions and infiltrate into the surrounding tissues. Malignancy is quite rare in these lesions. Common sites are head and neck, axilla, and mediastinum and rarely in breast. Hereby, we report a case of a 13-year-old female who presented with a gradually increasing painless swelling in the upper outer quadrant of her left breast for the last 12 years. Magnetic resonance imaging of the breast showed multiple, irregular, hypoechoic lesions associated with irregular duct dilatation. As the treatment of choice is complete excision, the cystic mass was excised in toto and sent for histopathology, which revealed cystic lymphangioma.

Keywords: *Breast, cystic lymphangioma, malignancy*

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Introduction

Cystic lymphangiomas or cystic hygromas consist of dilated lymph channels lined by an endothelium and are uncommon benign lymphatic tumor of the breast, particularly in adults. These occur due to the following reasons: blockage of the lymphatic channels with secondary dilatation, congenital weakness of the lymphatic wall, or proliferation of lymphatic vessels, resulting in lymph node degeneration, inflammation, and fibrosis of the existing lymph channels.[1] They are generally located in the head-and-neck area, the axilla, and mediastinum, and rarely in the retroperitoneum, abdominal organs, skeleton, pancreas, scrotum, or breast. Those involving the breasts are exceptionally uncommon, and a few cases have been reported in literature.[2] Lymphangiomas are almost universally benign tumors, though rare instances of malignant transformation have been reported.[3,4] Cystic lymphangiomas are most commonly diagnosed in young children; 50%–65% of lymphangiomas are clinically present in newborns and 90% are evident mostly by the age of 2 years.[5,6] Although generally a pediatric issue, cystic lymphangiomas can present in adults. Adult cystic lymphangiomas are also most prevalent in the neck, but have been observed in several different organ systems.[7] Cystic lymphangiomas of the breast have also been reported, though these are exceptionally rare. Cystic lymphangiomas are always congenital, but rarely acquired lymphangiomas of breast have been reported in adults as a late sequel of mastectomy and radiation therapy.[8]

Hereby, we report a case of congenital cystic lymphangioma of the left breast in a 13-year-old girl.

Case Report

A 13-year-old girl presented to the outdoor department with a complaint of a slow-growing, nontender mass in the left breast for the last 5 years. There was no associated nipple discharge or any other clinical symptoms. Physical examination revealed a nontender, slightly mobile, cystic mass in the upper, outer quadrant of the left breast measuring 20 cm × 15 cm. The overlying skin and areola were normal [Figure 1]. A clinical diagnosis of the cystic lesion of the breast was made. At first, fine-needle aspiration (FNA) cytology was performed, which was nonconclusive. Ultrasound showed a cyst in the upper and outer quadrants. Magnetic resonance imaging (MRI) was done to observe the

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extent of the cyst before excision [Figure 2]. Excision of the cyst was done after obtaining the consent of the patient [Figure 3]. A cystic structure measuring (20 cm × 15 cm) [Figure 4] was excised. Histopathology revealed large dilated spaces lined by a flattened endothelium growing in loose connective tissue as well as in glandular tissue of breast [Figure 5]. A final diagnosis of cystic lymphangioma of the breast was made. The diagnosis was made based on the clinical, radiological, and histopathological report.

Discussion

Cystic lymphangioma of the breast is a rare benign lymphatic tumor. It usually presents before the age of 2 years but may present later on in adolescence or adulthood, as in our case. The lymphangioma of breast, especially in adults, is an extremely unusual phenomenon. Most of the cystic lymphangiomas of the breast are mainly located in the upper outer quadrant of the breast, as in our case.[9,10] Rarely, cystic lymphangioma may be in male breast also.[11-13] Of the known cases, most are located in the upper, outer quadrant of the breast, as this area contains ~75% of the lymphatic drainage toward the tail and axilla, as in our case[14,15] Cystic lymphangioma is characterized by large, cyst-like spaces filled with clear lymph fluid and the spaces are lined by flat endothelial cells. Lymphangiomas can be classified into simple, cystic, and cavernous. Simple lymphangiomas consist of small, capillary-sized, thin-walled vessels with considerable connective tissue. Cystic lymphangiomas are characterized by a well-defined cyst such as spaces lined by endothelial cells and are filled with clear lymph fluid. Finally, cavernous lymphangiomas are composed of dilated lymphatic channels in a lymphatic stroma containing lymphoid aggregates.[16] Cystic lymphangioma is often associated with chromosomal abnormalities and other anatomical anomalies including Turner's syndrome, trisomy 21 and trisomy 18, as well as mosaic trisomy.[17]

The differential diagnosis of cystic lesions of the breast encompasses simple cysts, postoperative seromas, hematomas, lymphoceles, galactoceles, abscesses, hemangiomas, and, very rarely, lymphangiomas. Simple cysts are usually bilateral, and they often occur in other regions of the breast. They show a cyst with turbid yellow or greenish fluid without lining cells. Hemangiomas generally appear with thick-walled blood vessels with many red blood cells.[10,18]

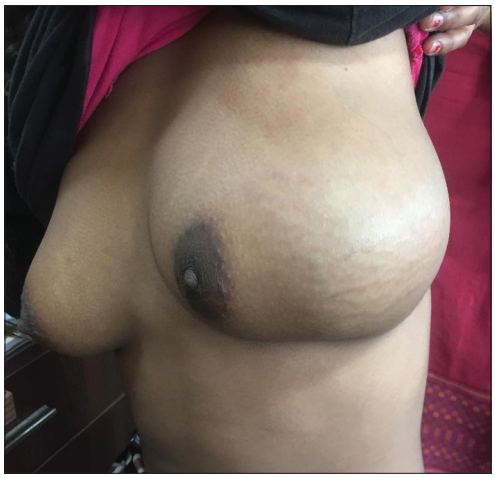

Figure 1: Patient's image: swelling in the left breast

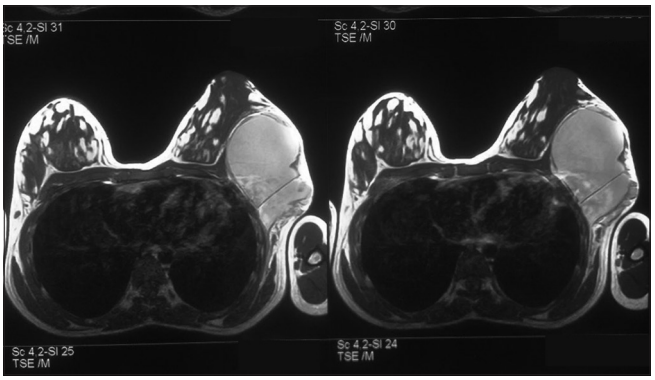

Figure 2: Magnetic resonance imaging of the breast

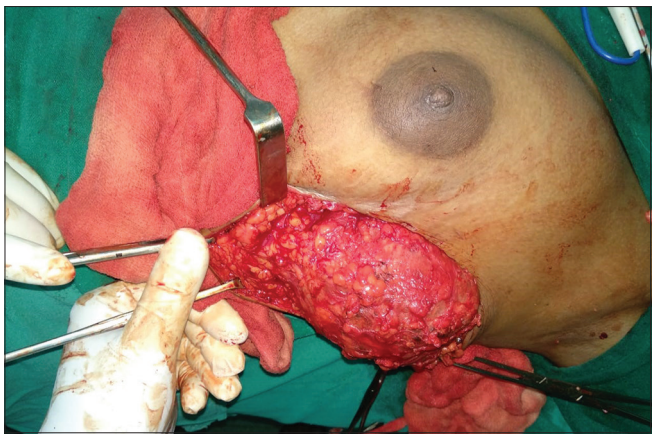

Figure 3: Operative part

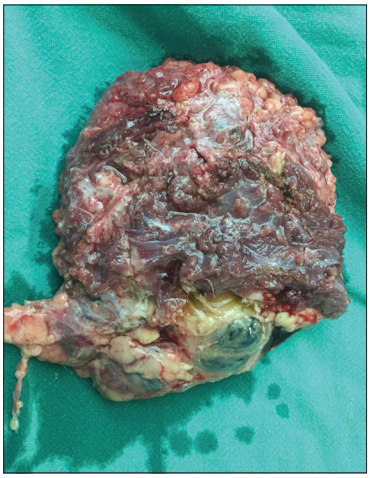

Figure 4: Excised cyst

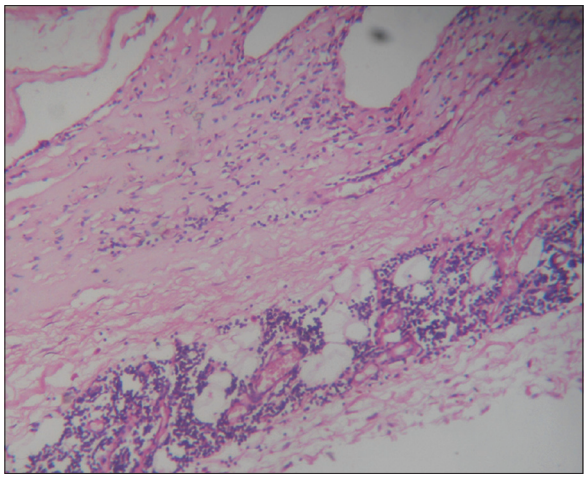

Figure 5: Histopathology report

Mammography is usually nonspecific revealing fairly or ill-defined, round or lobulated densities interspersed between the normal breast parenchyma without obvious signs of micro or macrocalcification. In many cases, axillary and posterior forbidden areas may be unreachable due to macromastia.

Ultrasonography may reveal dilated noncompressible lymphatic ducts joining toward the axillary region, appearing anechoic or hypoechoic secondary to proteinaceous or hemorrhagic fluid. In early cases, cystic areas may mimic fibrocystic disease of breast, which is usually bilateral or hemangiomatous lesion, which shows vascularity on power Doppler imaging.

MRI is the modality of choice for evaluating breast lymphangioma. It helps in differentiation between benign and malignant as well as determines the extent of swelling. Breast lymphangioma appears as cystic septate masses with variably ductal dilatation and only septal enhancement, with some of the ducts/cysts showing hyperintense content on both T1-weighted and T2-weighted images due to proteinaceous or hemorrhagic contents, as seen in our case.[12,17] Immunohistochemical investigations can distinguish between hemangioma and lymphangioma.[18]

About 31 cases of mammary cystic lymphangiomas have been reported in literature during the period of 2003–2019 [Table 1]. The reason for their late onset is not clear in all reports, and no particularly related history of pregnancy or breastfeeding was identified except in a report by de Guerké *et al*.[9]

As these tumors are usually present at birth, careful history taking is very significant. Delay in diagnosis is associated with increase in the size of tumor, and also there may be invasion and mixing with normal breast tissue and thus may produce the heterogeneous pattern of tumor and macromastia.[24,25,31,35] The definitive diagnosis, however, can be established by core or excision biopsy. Histopathology is confirmatory for final diagnosis.[14] There was no history of breast surgery, trauma, or infection in our case. The lymphatic endothelial lining cells are strongly positive for D240 and negative for CD34 (endothelial marker for blood vessels) on immunohistochemical staining. This test was not done as this facility does not exist in our institution.[36]

Table 1: Mammary Cystic Lymphangioma (Period 2003-2019).

Author	Sex	Age (years)	Size (cm)	Side
Chung *et al*., 2003[15]	F	34	10	Right
Waqar *et al*., 2004[10]	F	24	20×7	Right
Yaghan *et al*., 2004[19]	M	30		Right
de Guerké *et al*., 2005[9]	F	31	3	Left
Torcasio *et al*., 2006[4]	F	26	4×3×2	Right
Krainick-Strobel *et al*., 2006[18]	F	43	15×10	Left
Ogun *et al*., 2007[14]	F	38	5×4	Left
Min *et al*., 2008[20]	F	36	4×3	right
Kwon *et al*., 2009[21]	F	31	20×17	Left
Singh *et al*., 2009[12]	M	6	4.5×3.5×2.5	Left
Ko *et al*., 2009[37]	F	34	1.5	Left
Sasi *et al*., 2010[39]	F	37	10	Left
Balaji and Ramachandran, 2010[17]	F	23	-	Left
Malhotra *et al*., 2010[23]	M	60	7×7×5	Left
Hahn *et al*., 2011[5]	F	51	---	right
Nguyen *et al*., 2011[24]	F	71	6	Left
Ekmez *et al*., 2011[11]	M	05	6×4	left
Gupta and Singh, 2011[13]	M	8	7×6.5×3	Right
Hynes *et al*., 2012[25]	F	33	-	Left
Harbade *et al*., 2013[26]	F	23	20×10	Right
Hiremath and Binu, 2014[27]	F	23	6×6×7	Right
Alkhalili *et al*., 2014[28]	F	47	0.7	left
Vargas-Hernández *et al*., 2014[29]	F	45	-	Left
Arafah *et al*., 2015[30]	F	37	20×15×10	left
Rusdianto *et al*., 2016[16]	F	20	3×1.5×1.5	Left
Rastogi, 2016[31]	F	16	4.5×3	right
Almohawes *et al*., 2017[32]	F	39	-	Right
Chotai *et al*., 2018[33]	F	41	3.9×3.6	Left
Park *et al*., 2018[34]	F	37	3.3	Right
Principe *et al*., 2019[2]	F	27	20×10	left
Present case	F	13	20×15	Left

FNA fluid aspirates always reveal clear, yellowish, or straw-colored fluid, with plenty of lymphocytes. However, FNA is usually not diagnostic as rarely shows cells. Therefore, surgical excision with histopathological diagnosis is the mainstay of diagnosis.[30,37]

Lymphangiomas are treated for cosmetic and functional reasons. The choice of treatment depends on the size, depth, and location of the lesion. Undoubtedly, patient's age and general health are also important. Sclerosing agents such as steroid or bleomycin injections cause fibrosis, making surgery difficult and if used as monotherapy, there are high chances of recurrence.[38] However, for patients in whom surgery is contraindicated, guided percutaneous sclerosis can be performed.

Surgical excision is the mainstay of treatment. Ideal is wide surgical excision of the cisterns, but there may be difficulty in complete excision due to the tendency of these lesions to infiltrate into the surrounding normal breast tissues. In our case, wide excision was possible as there was no infiltration in the surrounding tissues. Incomplete excision always leads to rapid recurrence. Streptococcal lysine (OK-432), a biological response modifier, causing shrinkage of cystic spaces, can be used for small residual or recurred lymphangioma.[38]

Recurrence can occur following incomplete excision. As reported, recurrence rates of cystic lymphangiomas in pediatric groups are 100% after aspiration, 100% after injection, 40% after incomplete excision, 40% after laser treatment, and 17% after macroscopic total excision. Therefore, in mammary lesions, the goal of treatment should be preferably a complete excision to reduce the recurrence risk.[39]

Conclusion

We report here a rare case of cystic lymphangioma of the breast in a 13-year-old girl. Although the incidence of this disease is extremely rare in adult breast, cystic lymphangioma should be considered in the differential diagnosis of cystic mass in the upper and outer quadrants of the breast. Early recognition of this rare disease is of great importance because complete surgical excision of small lesions and consequent cure is possible.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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