

# Concomitant breast and axillary lymphangioma in an adult

## A case report and a review of the literature

Taejin Park, MD<sup>a</sup>, Han Shin Lee, MD<sup>a</sup>, Eun Jung Jung, MD<sup>a,\*</sup>, Ju Yeon Kim, MD<sup>b</sup>, Chi Young Jeong, MD<sup>b</sup>, Young Tae Ju, MD<sup>b</sup>, Young Joon Lee, MD<sup>b</sup>, Soon Chan Hong, MD<sup>b</sup>, Bo Hwa Choi, MD<sup>c</sup>, Hyo Jung An, MD<sup>d</sup>

### Abstract

**Rationale:** Lymphangiomas develop in the head, neck, and axilla of patients <2 years old in more than 90% of cases. They are rarely reported in adults.

**Patient concerns:** Here, we report on a 37-year-old woman with a firm, hypoechoic 3.3 cm mass in the right upper, outer quadrant of the breast with discomfort, and swelling of the right axillary region.

**Diagnosis and interventions:** She underwent wide excision of the right breast and axillary lesion and the lesion pathologic finding is lymphangioma of the breast.

**Outcomes:** She was in good condition with no signs of postoperative complications and no evidence of recurrence at 6 months postsurgery.

**Lessons:** Despite the rarity of breast cystic lymphangioma, its evaluation should be considered for prompt diagnosis and definitive treatment to prevent recurrence and complications. Furthermore, this is the first case of concomitant lymphangioma of the breast parenchyma and axillary region.

**Keywords:** adult, axillary, breast, lymphangioma

## 1. Introduction

Lymphangioma is a benign lymphatic tumor typically found in children; 90% of the cases are diagnosed before 2 years of age.<sup>[1,2]</sup> Most occurrence regions of lymphangioma are the head and neck area (75%) and the axilla (20%).<sup>[3,4]</sup> Lymphangioma of the breast is very rare in adults, and a few cases have been reported.<sup>[5]</sup> Breast lymphangiomas are mainly located in the subareolar and upper, outer quadrant of the breast.<sup>[5,6]</sup> Complete surgical excision is the preferred treatment.<sup>[6]</sup> We report a unique case of concomitant lymphangioma of the axillary region and upper, outer quadrant of the breast.

Editor: N/A.

The authors report no conflicts of interest.

<sup>a</sup> Department of Surgery, Gyeongsang National University School of Medicine and Gyeongsang National University Changwon Hospital, Changwon,

<sup>b</sup> Department of Surgery, Gyeongsang National University School of Medicine and Gyeongsang National University Hospital, Jinju, <sup>c</sup> Department of Pathology,

<sup>d</sup> Department of Radiology, Gyeongsang National University School of Medicine and Gyeongsang National University Changwon Hospital, Changwon, Republic of Korea.

\* Correspondence: Eun Jung Jung, Department of Surgery, Gyeongsang National University School of Medicine and Gyeongsang National University Changwon Hospital, Changwon, Republic of Korea (e-mail: drfej@gnu.ac.kr).

Copyright © 2018 the Author(s). Published by Wolters Kluwer Health, Inc. This is an open access article distributed under the terms of the Creative Commons Attribution-Non Commercial-No Derivatives License 4.0 (CCBY-NC-ND), where it is permissible to download and share the work provided it is properly cited. The work cannot be changed in any way or used commercially without permission from the journal.

Medicine (2018) 97:45(e12946)

Received: 2 May 2018 / Accepted: 28 September 2018

<http://dx.doi.org/10.1097/MD.00000000000012946>

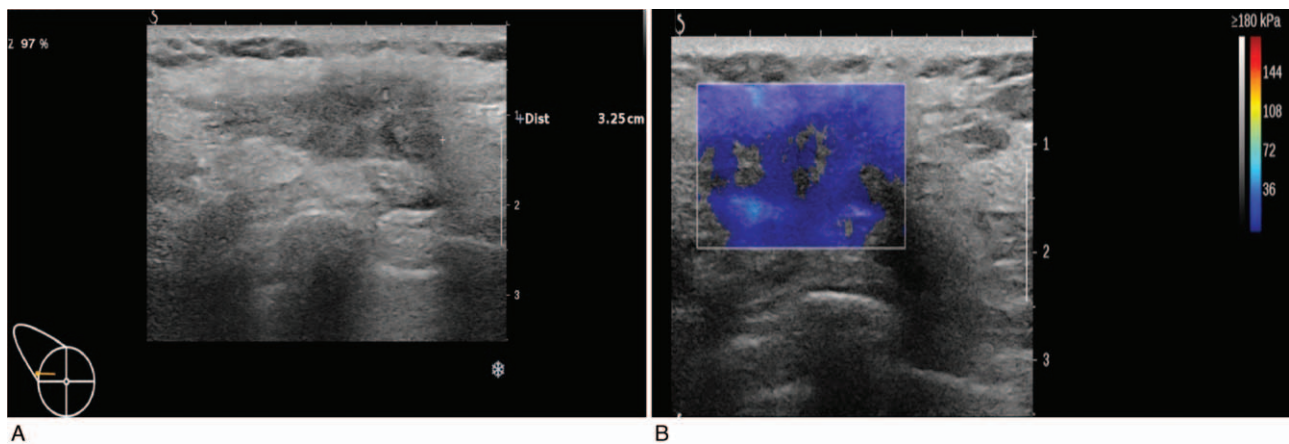
## 2. Methods

Because this case report is not a prospective or retrospective study, the consent of the patient was sufficient, and ethical approval was not required. Thus, we decided to publish only the age, image findings, and pathologic pictures in the case report, and we received written consent from the patient.

## 3. Case report

### 3.1. Clinical summary

A 37-year-old woman with no medical history visited our department with a mass in her right breast. Three weeks before her visit, she went to another hospital because of her right breast mass and was clinically diagnosed with hemangioma by breast ultrasound. Upon physical examinations, a mild asymmetry was detected in her right breast, and there was a firm, static mass on the right upper, outer quadrant. The ultrasound revealed a 3.3 cm indistinct, hypoechoic lesion at the right 10 o'clock position, which was classified using the Breast Imaging-Report and Data System 4A (Fig. 1). The right side was more prominent upon examination of both axillary accessory breast tissues. Ultrasound-guided core needle biopsy was performed, and lymphovascular lesions were detected; the patient had discomfort and swelling of the right axilla following the core needle biopsy and subsequently underwent a wide excision of the right breast. In the surgical field, the mass was found to be hard and nonmovable, and a duct was leading toward the axillary direction. The axillary cystic structures in the axillary area had communication with the duct. The axillary cystic structures were identified and consisted of multiple thin and fibrotic septae filled with a clear fluid (Fig. 2). Following the excision, the patient has shown no signs of



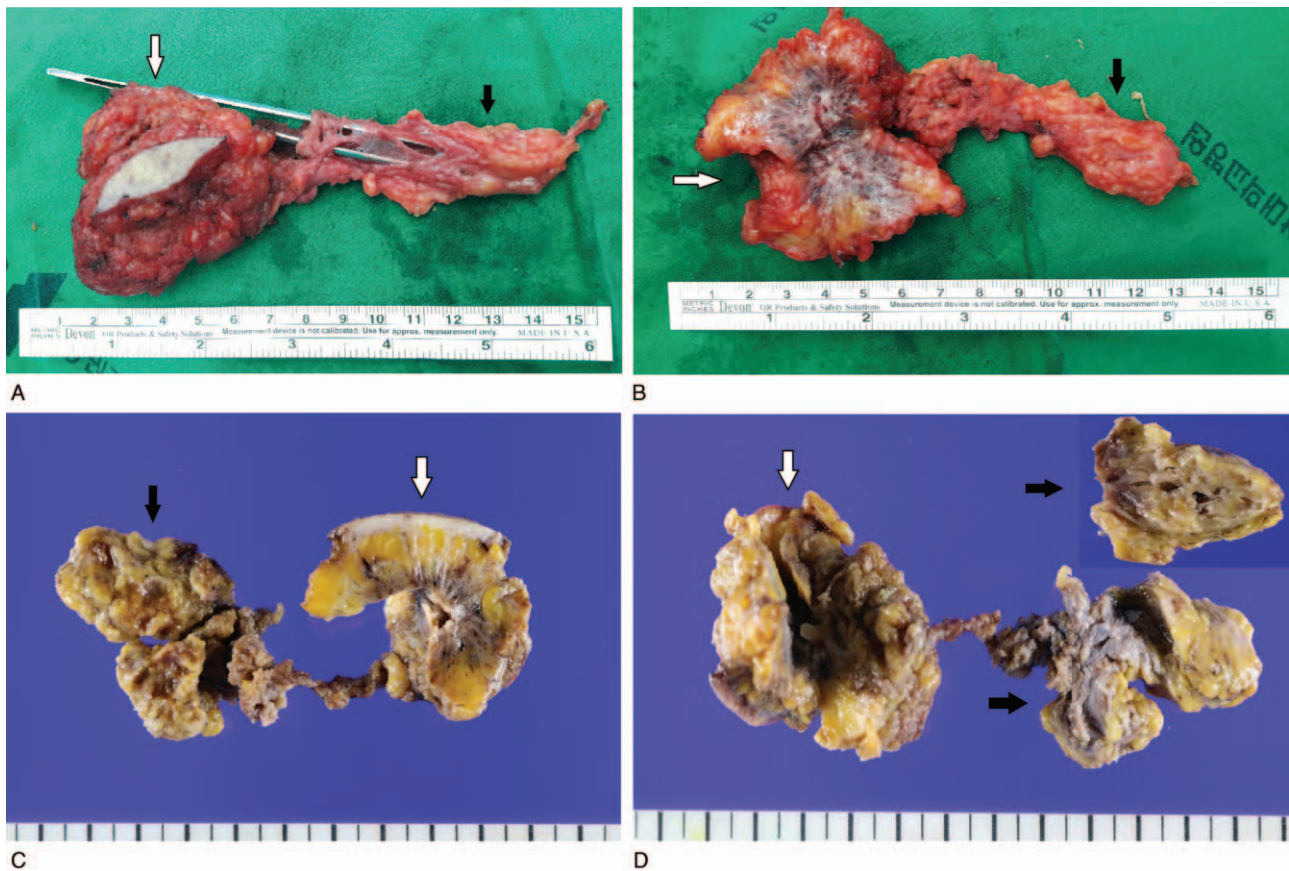
**Figure 1.** A, Ultrasonography identified a 3.3cm hypoechoic, irregular lesion. B, Doppler image.

postoperative complications and no evidence of recurrence at 6 months postsurgery.

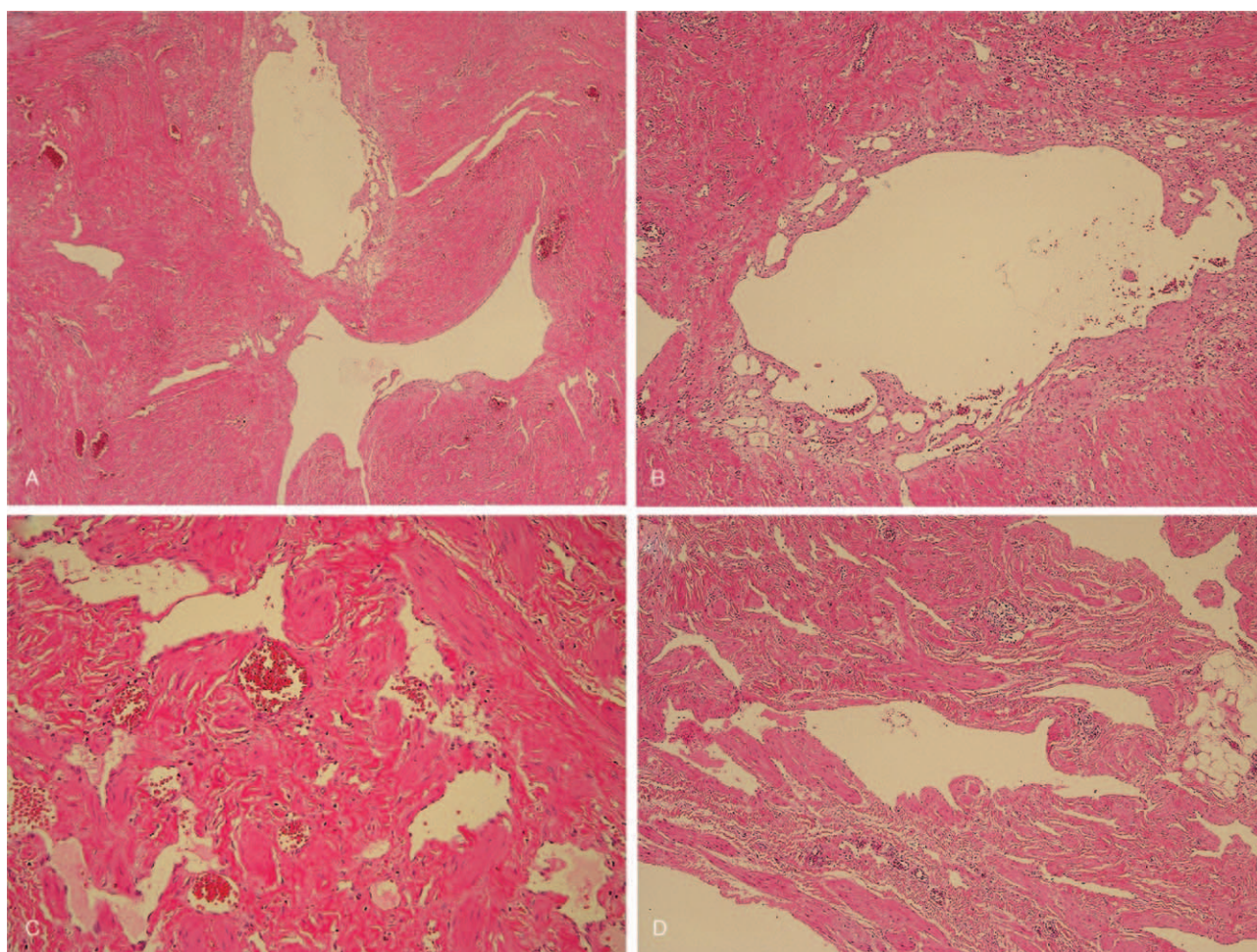
### 3.2. Pathological findings

The breast lesion was hard and grayish white, with irregular margins. There was a duct leading toward the right axillary

direction which was connected to another lesion in the axillary area (Fig. 2 A, B). The axillary lesion was composed of multiple thin septae filled with a clear fluid. Cystic cavities were observed in the center of the breast lesion (Fig. 2 C, D). The final diagnosis was concomitant breast and axillary lymphangioma (Fig. 3). The immunohistochemical results were as follows: calponin-1 (-); CD-31 (+); D2-40 (+); Ki-67 (-) (Fig. 4).



**Figure 2.** Macroscopic findings. A, Right breast mass measuring  $5.5 \times 4.0 \times 4.5$  cm. B, Right axillary mass measuring  $5.0 \times 3.0 \times 1.2$  cm. The right breast mass was hard and grayish white with irregular margins; the right axillary mass had multiloculated thin septae containing a clear yellowish fluid. C, The center of the breast mass consisted of cystic cavities. D, Close-up of axillary mass specimen showing the cystic septae (white arrows; breast mass, black arrows; axillary mass).

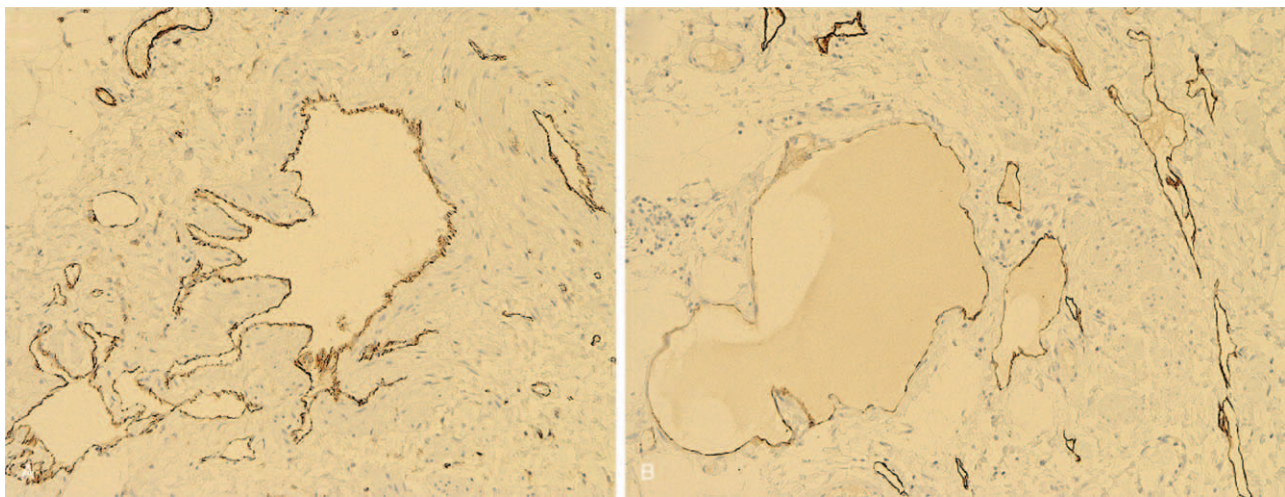


**Figure 3.** Microscopic findings. A, Dilated empty vascular channels extending through the fibrous stroma in the right breast mass [hematoxylin and eosin (H&E),  $\times 10$ ]. B, The stroma surrounding the vascular channels contained sparse smooth muscle cells and lymphocytes in the right breast mass (H&E,  $\times 40$ ). C, Under higher magnification, dilated blood-filled and empty vascular channels were detected. The flat endothelium was inconspicuous in the right breast mass (H&E,  $\times 100$ ). D, In the right axillary mass, irregularly shaped vascular channels similar to those in the tumor of the breast are seen (H&E,  $\times 100$ ).

#### 4. Discussion

Lymphangiomas are rare benign lymphatic malformations resulting from lymphatic dilatation with endothelial linings. They are a slow-growing, benign tumor<sup>[1]</sup> caused by congenital weakness of the wall, blockage of the lymphatic channels, or proliferation of lymphatic vessels.<sup>[7]</sup> Initially, they may be diagnosed as simple cysts, lymphoceles, hematomas, or heman-giomas.<sup>[8]</sup> Lymphangiomas are simple, cystic, and cavernous according to their pathologic features.<sup>[9]</sup> Simple lymphangiomas consist of small-sized capillaries and thin-walled vessels. Cystic lymphangiomas consist of spaces with well-defined cysts lined by endothelial cells, filled with a clear fluid. A cavernous lymphangioma consists of dilated lymphatic channels containing lymphoid aggregates.<sup>[9]</sup> More than 70% of lymphangiomas occur in the neck; 20% occur in the axillary region, and 10% occur in the abdomen, skeleton or scrotum.<sup>[3]</sup> Breast lymphangiomas are very rare with  $<20$  cases reported in the last 4 decades.<sup>[5]</sup> Thirty-five cases of mammary cystic lymphangiomas have been reported worldwide in the last 50 years (Table 1). Thirty cases occurred in females aged 16 to 71 years, and 5 cases were reported in men. The size of the mammary lymphangioma ranges from 0.7 to 25 cm, and the most common sites are the

upper, outer quadrant, and subareolar region.<sup>[5]</sup> This may be related to the lymphatic drainage of the breast; the axillary lymph nodes receive  $>75\%$  of the fluid drained from the lateral quadrants of the breast.<sup>[38]</sup> In this case, the location of the breast lymphangiomas was the right axillary region and the right upper, outer quadrant, connected by a lymphatic duct. Although the axillary lymphangioma was a typical cystic lymphangioma, the breast lymphangioma had undetermined features resembling a malignant tumor. The cross-section of the breast mass, however, revealed cystic lymphangiomas (Fig. 2). Thus, we discussed about the potential mechanism of this concomitant breast and axillary lymphangioma. First, we suspected that the patient had right axillary lymphangiomas that were not detected in childhood. Second, the axillary lymphangioma extended to the upper, outer quadrant with development of a lymphatic channel in the breast. In the operation field, the breast lymphangiomas had a little fluid collection and collapsed remnant cystic lesions. Likewise, the axillary lymphangioma had multiple, thin, and fibrotic septae filled with a small amount fluid. For this reason, the axillary lymphangioma was not detected by ultrasound. The mammography identified a round/lobulated lesion with increased opacity, whereas the ultrasonography showed a multiloculated, hypo-echoic, cystic mass, with variable sized septa with solid



**Figure 4.** Immunohistochemical findings. The vascular channels show positive expression of the lymphovascular markers CD31 (A,  $\times 100$ ) and D2-40 (B,  $\times 100$ ).

components.<sup>[31]</sup> Computed tomography (CT) or magnetic resonance imaging (MRI) provides more accurate images and a more in-depth assessment of the tumor.<sup>[10]</sup> In MRI, cystic lymphangiomas are seen as septated masses with a low T1-

weighted and high T2-weighted signal intensity.<sup>[28]</sup> In this case, the ultrasound showed an indistinct, hypoechoic mass, and no axillary masses. Consequently, we performed a right breast mass excision, and the right axillary cystic lymphangioma was

**Table 1**

**Case reports of cystic lymphangioma in the breast and axillary region.**

No	Author	Age, y	Sex	Size, cm	Location	Management	Country	Year
1	Hessler et al <sup>[10]</sup>	28	F	7	Rt. areola	Surgery	Sweden	1967
2	Pace and Schrivere <sup>[11]</sup>	44	F	10	Lt. breast	Surgery	Italy	1967
3	Sieber and Sharkey <sup>[12]</sup>	49	F	7 $\times$ 7 $\times$ 2	Lt. upper outer quadrant	Surgery	USA	1986
4	Salvador et al <sup>[8]</sup>	19	F	10 $\times$ 9	Rt. breast	Surgery	Spain	1990
5	Kangesu <sup>[13]</sup>	6	M	2	Lt. whole breast	Surgery	England	1990
6	Kurosumi et al <sup>[14]</sup>	16	F	16 $\times$ 14	Rt. breast	Surgery	Japan	1991
7	Tolpinski and Bakhaev <sup>[15]</sup>	—	F	—	—	Surgery	Russia	1992
8	Meunier et al <sup>[16]</sup>	30	F	12 $\times$ 4	Rt. areola lower part	Surgery	France	1994
9	Chiba and Ibrahim <sup>[17]</sup>	4 mo	M	—	Lt. whole breast	Surgery	Japan	1995
10	Occhiato et al <sup>[18]</sup>	27	F	2.5	Lt. breast	Surgery	Italy	1996
11	Sa and Choi <sup>[3]</sup>	36	F	3.5 $\times$ 3	Lt. upper outer quadrant	Surgery	Korea	1999
12	Aryya et al <sup>[19]</sup>	35	F	18 $\times$ 15	Rt. whole breast	Surgery	India	1999
13	Chung et al <sup>[20]</sup>	34	F	10	Rt. upper outer quadrant	Surgery	Korea	2003
14	Waqar et al <sup>[21]</sup>	24	F	25 $\times$ 20	Rt. diffuse breast	Surgery	Afghanistan	2004
15	Yaghan and Bani-Hani <sup>[22]</sup>	30	M	—	Rt. diffuse breast	Surgery	Jordan	2004
16	De Guerke et al <sup>[23]</sup>	31	F	3	Lt. upper outer quadrant	Conservative	France	2005
17	Krainick-Strobel et al <sup>[24]</sup>	43	F	15 $\times$ 10	Lt. upper outer quadrant	Surgery	Germany	2006
18	Torcasio et al <sup>[25]</sup>	26	F	3	Rt. inner quadrant	Surgery	Italy	2006
19	Ogun et al <sup>[6]</sup>	38	F	5 $\times$ 4	Lt. upper outer quadrant	Surgery	Nigeria	2007
20	Min et al <sup>[26]</sup>	36	F	4 $\times$ 3	Rt. upper outer quadrant	Surgery	Korea	2008
21	Kwon et al <sup>[2]</sup>	31	F	20 $\times$ 17	Lt. whole breast	Surgery	Korea	2009
22	Sasi et al <sup>[27]</sup>	37	F	10	Lt. upper outer quadrant	Surgery	England	2010
23	Balaji and Ramachandran <sup>[28]</sup>	23	F	—	Lt. whole breast	—	India	2010
24	Malhotra et al <sup>[29]</sup>	60	M	7 $\times$ 7 $\times$ 5	Lt. upper outer quadrant	Surgery	India	2010
25	Nguyen et al <sup>[30]</sup>	71	F	6	Lt. axillary	Surgery	USA	2011
26	Gupta and Singh <sup>[31]</sup>	8	M	7 $\times$ 6.5 $\times$ 3	Rt. upper outer quadrant	Surgery	India	2011
27	Hynes et al <sup>[32]</sup>	33	F	—	Lt. whole breast	Surgery	Ireland	2012
28	Harbade et al <sup>[33]</sup>	23	F	20 $\times$ 10	Rt. upper outer quadrant	Surgery	India	2013
29	Hiremath and Binu <sup>[7]</sup>	23	F	6 $\times$ 6 $\times$ 7	Rt. areola lower	Surgery	India	2014
30	Alkhalili et al <sup>[34]</sup>	47	F	0.7	Lt. nipple	Punch biopsy	USA	2014
31	Vargas-Hernandez et al <sup>[35]</sup>	45	F	—	Lt. whole breast	Surgery	Mexico	2014
32	Arafah et al <sup>[36]</sup>	37	F	20 $\times$ 15 $\times$ 10	Lt. upper outer quadrant	Surgery	USA	2015
33	Rusdianto et al <sup>[5]</sup>	20	F	3 $\times$ 1.5 $\times$ 1.5	Lt. inner outer quadrant	Surgery	USA	2016
34	Almohawes et al <sup>[37]</sup>	39	F	—	Rt. whole breast	Surgery	Saudi Arabia	2017
35	Chotai et al <sup>[8]</sup>	41	F	3.9 $\times$ 3.6	Lt. axillary	Surgery	Singapore	2017

discovered during the surgery. We considered doing an MRI for further evaluation of the breast mass; however, it was not possible due to the high cost and problems with the patient's insurance. Immunohistochemical investigations are able to distinguish between hemangioma and lymphangioma.<sup>[24]</sup> Lymphatic endothelial markers are CD31, CD34, podoplanin, LYVE-1, and PORX-1. Otherwise, the vascular endothelial marker is VIII-associated antigen.<sup>[14]</sup> In our case, the vascular channels showed a positive expression of CD31 and D2-40 (Fig. 4).

The treatment of choice for breast lymphangioma is complete surgical excision. Different options include incision and drainage, sclerotherapy, steroid, radiotherapy, and carbon dioxide laser.<sup>[21]</sup> However, these are associated with high rate of recurrence.<sup>[7]</sup> So complete surgical excision is needed for low probability of recurrence.<sup>[6]</sup> Spontaneous resolution is uncommon.<sup>[23]</sup> Among the cases identified in the literature, almost all breast lymphangiomas were removed surgically, and only 1 case was treated conservatively (Table 1).

## 5. Conclusions

To our knowledge, this is the first case of concomitant lymphangioma of the breast parenchyma and axillary region. Despite its rarity, evaluation for breast cystic lymphangioma should be considered for a prompt diagnosis and definitive treatment to prevent recurrence and complications.<sup>[21]</sup> Complete surgical excision is the most effective option.

## Author contributions

**Conceptualization:** Eun Jung Jung, Taejin Park.

**Data curation:** Taejin Park, Han Shin Lee, Bo Hwa Choi, Hyo Jung An.

**Formal analysis:** Taejin Park, Han Shin Lee, Eun Jung Jung.

**Investigation:** Taejin Park.

**Supervision:** Eun Jung Jung.

**Writing – original draft:** Taejin Park.

**Writing – review and editing:** Eun Jung Jung, Ju Yeoun Kim, Chi Young Jeoung, Young Tae Ju, Young Joon Lee, Soon Chan Hong, Bo Hwa Choi, Hyo Jung An.

## References

- [1] Hahn SH, Choi HY, Park SH, et al. Lymphangioma and lymphangiectasia of the breast mimicking inflammatory breast cancer. *J Ultrasound Med* 2011;30:863–5.
- [2] Kwon SS, Kim SJ, Kim L, et al. Huge cystic lymphangioma involving the entire breast. *Ann Plast Surg* 2009;62:18–21.
- [3] Sa EJ, Choi YH. Cystic lymphangioma of the breast. *J Clin Ultrasound* 1999;27:351–2.
- [4] Chotai N, Fok E, Chan P, et al. Axillary lymphangioma in an asymptomatic adult female. *Breast J* 2018;24:415–6.
- [5] Rusdianto E, Murray M, Davis J, et al. Adult cystic lymphangioma in the inner quadrant of the breast-rare location for a rare disease: a case report. *Int J Surg Case Rep* 2016;20:123–6.
- [6] Ogun GO, Oyetunde O, Akang EE. Cavernous lymphangioma of the breast. *World J of Sur Oncol* 2007;5:69.
- [7] Hiremath B, Binu V. Case Report: Lymphangioma of the breast. *BMJ Case Reports* 2014;2014 Mar 17.
- [8] Salvador R, Salvador M, Miranda D, et al. Cystic hygroma of the breast. *Eur J Radiol* 1990;11:215–7.
- [9] Kumar V, Abbas AK, Fausto N. Robbins & Cotran Pathologic Basis of Disease. 7th ed 2005;WB Saunders, Philadelphia, PA:547.
- [10] Hessler C. Cystic lymphangioma of the breast. *Radiology* 1967;88:135–7.
- [11] Pace M, Schrivere G. Cystic lymphangioma of the breast in adults. *Friuli Med* 1967;22:561–70.
- [12] Sieber PR, Sharkey FE. Cystic hygroma of the breast. *Arch Path Lab Med* 1986;110:353.
- [13] Kangesu T. Cystic hygroma of the breast in childhood. *Br J Clin Pract* 1990;44:787–8.
- [14] Kurosumi M, Namoto C, Suemasu K, et al. Cavernous lymphangioma of the breast: case report with electron microscopic and immunohistochemical investigation. *Jpn J Clin Oncol* 1991;21:129–34.
- [15] Tolpinski AP, Bakhlaev IE. Giant lymphoma of the breast. *Vopr Onkol* 1992;38:233–4.
- [16] Meunier L, Barneon G, Meynadier J. Acquired progressive lymphangioma. *Br J Dermatol* 1994;131:706–8.
- [17] Chiba T, Ibrahim M. Cavernous lymphangioma of the breast: case report of an infant. *Nippon Geka Hokan* 1995;64:23–6.
- [18] Occhiato R, Russo F, Broglia L. Simple lymphangioma of the breast. *Radiol Med (Torino)* 1996;91:130–2.
- [19] Aryya NC, Rastogi AN, Kumar K, et al. Giant lymphangioma of the breast in an adult female. *Aust N Z J Surg* 1999;69:891–2.
- [20] Chung SY, Oh KK, Kim DJ. Mammographic and sonographic findings of a breast cystic lymphangioma. *J Ultrasound Med* 2003;22:307–9.
- [21] Waqar SN, Khan H, Mekan SF, et al. Cystic breast lymphangioma. *J Pak Med Assoc* 2004;54:531–3.
- [22] Yaghan RJ, Bani-Hani KM. Male breast disorders in Jordan. Disease patterns and management problems. *Saudi Med J* 2004;25:1877–83.
- [23] De Guerké L, Baron M, Dessogne P, et al. Cystic lymphangioma of the breast. *Breast J* 2005;11:515–6.
- [24] Krainick-Strobel U, Kramer B, Walz-Mattmuller R, et al. Massive cavernous lymphangioma of the breast and thoracic wall: case report and literature review. *Lymphology* 2006;39:146–51.
- [25] Torcasio A, Veneroso S, Amabile MI. Cystic hygroma of the breast: a rare lesion. *Tumori* 2006;92:347–50.
- [26] Min KW, Jang SH, Na W, et al. Cystic lymphangioma of the breast in an adult woman. *Korean J Pathol* 2008;42:244–6.
- [27] Sasi W, Schneider C, Shah R, et al. Recurrent cystic lymphangioma of the breast: case report and literature review. *Breast Dis* 2010;31:43–7.
- [28] Balaji R, Ramachandran K. Cystic lymphangioma of the breast: magnetic resonance imaging features. *Breast Care (Basel)* 2010;5:250–2.
- [29] Malhotra P, Bansal A, Chintamani, et al. Cavernous lymphangioma of the male breast. *Indian J Pathol Microbiol* 2010;53:853–4.
- [30] Nguyen K, Karsif K, Lee S, et al. Lymphangioma in an elderly patient: an unusual cause of axillary mass. *Breast J* 2011;17:416–7.
- [31] 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;41:1314–8.
- [32] Hynes SO, McLaughlin R, Kerin M, et al. A unique cause of a rare disorder, unilateral macromastia due to lymphangiomatosis of the breast: a case report. *Breast J* 2012;18:367–70.
- [33] Harbade SR, Wasadikar PP, Varudkar AS. Giant lymphangioma of the breast in an adult. *J Evolution Med Dent Sci* 2013;2:3257–62.
- [34] Alkhalili E, Ayoubieh H, O'Brien W, Billings SD. Case Report: Acquired progressive lymphangioma of the nipple. *BMJ Case Rep* 2014;2014 Sep 22.
- [35] Vargas-Hernandez VM, Tovar-Rodríguez JM, Mario Adan M-E, Acosta-Altamirano G. Giant cystic lymphangioma breast: report of a case with 20-year follow-up and review of the literature. *Cir Cir* 2014;82:81–6.
- [36] Arafah M, Sweet G, Ginter PS, et al. Mammary lymphangioma. *Int J Surg Pathol* 2015;23:542–3.
- [37] Almohawes E, Khoumais N, Arafah M, et al. Cystic lymphangioma of the breast: a case report in an adult woman. *OMICS J Radiol* 2017;6:2.
- [38] Hultborn K, Larsson L-G, Ragnhult I. The lymph drainage from the breast to the axillary and parasternal lymph nodes, studied with the aid of colloidal AUI98. *Acta Radiologica* 1955;43:52–64.