Concomitant breast and axillary lymphangioma in an adult
A case report and a review of the literature

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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 findings 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.[1,2] Most occurrence regions of lymphangioma are the head and neck area (75%) and the axilla (20%).[3,4] Lymphangioma of the breast is very rare in adults, and a few cases have been reported.[5] Breast lymphangiomas are mainly located in the subareolar and upper, outer quadrant of the breast.[6] Complete surgical excision is the preferred treatment.[6] We report a unique case of concomitant lymphangioma of the axillary region and upper, outer quadrant of the breast.

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...
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 × 4.0 × 4.5 cm. B, Right axillary mass measuring 5.0 × 3.0 × 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).
4. Discussion

Lymphangiomas are rare benign lymphatic malformations resulting from lymphatic dilatation with endothelial linings. They are a slow-growing, benign tumor caused by congenital weakness of the wall, blockage of the lymphatic channels, or proliferation of lymphatic vessels. Initially, they may be diagnosed as simple cysts, lymphoceles, hematomas, or hemangiomas. Lymphangiomas are simple, cystic, and cavernous according to their pathologic features. 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. More than 70% of lymphangiomas occur in the neck; 20% occur in the axillary region, and 10% occur in the abdomen, skeleton or scrotum. Breast lymphangiomas are very rare with <20 cases reported in the last 4 decades. 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. 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. 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 lymphangioma 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, hypoechoic, cystic mass, with variable sized septa with solid...
Computed tomography (CT) or magnetic resonance imaging (MRI) provides more accurate images and a more in-depth assessment of the tumor. In MRI, cystic lymphangiomas are seen as septated masses with a low T1-weighted and high T2-weighted signal intensity. 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 excised.

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[10]    | 28     | F   | 7        | Rt. areola                | Surgery      | Sweden       | 1967 |
| 2  | Pace and Schrivere[11]| 44     | F   | 10       | Lt. breast                | Surgery      | Italy         | 1967 |
| 3  | Sieber and Sharkey[2] | 49     | F   | 7 x 7 x 2| Lt. upper outer quadrant  | Surgery      | Spain         | 1990 |
| 4  | Salvador et al[8]    | 19     | F   | 10 x 9   | Rt. breast                | Surgery      | England       | 1990 |
| 5  | Kangesu[3]           | 6      | M   | 2        | Lt. whole breast          | Surgery      | Japan         | 1991 |
| 6  | Kurosumi et al[4]    | 16     | F   | 16 x 14  | Rt. breast                | Surgery      | Japan         | 1991 |
| 7  | Topilinski and Bakhtlaev[5] | —     | F   | —        | Lt. areola lower part     | Surgery      | Russia        | 1992 |
| 8  | Mounier et al[6]     | 30     | F   | 12 x 4   | Lt. whole breast          | Surgery      | France        | 1994 |
| 9  | Chiba and Ibrahim[7] | 4 mo   | M   | —        | Lt. breast                | Surgery      | Japan         | 1995 |
| 10 | Occhiato et al[8]    | 27     | F   | 2.5      | Lt. upper outer quadrant  | Surgery      | Korea         | 1999 |
| 11 | Sa and Choi[9]       | 36     | F   | 3.5 x 3  | Rt. whole breast          | Surgery      | India         | 1999 |
| 12 | Aryya et al[10]      | 35     | F   | 18 x 15  | Rt. upper outer quadrant  | Surgery      | Korea         | 2003 |
| 13 | Chung et al[11]      | 34     | F   | 10       | Rt. whole breast          | Surgery      | Afghanistan   | 2004 |
| 14 | Waqar et al[12]      | 24     | F   | 25 x 20  | Rt. whole breast          | Surgery      | Jordan        | 2004 |
| 15 | Yaghan and Bani-Harb[13]| 30      | M   | —        | Lt. upper outer quadrant  | Conservative  | France        | 2006 |
| 16 | De Guerke et al[14]  | 31     | F   | 3        | Lt. whole breast          | —            | —            | 2010 |
| 17 | Kainzick-Strobel et al[15]| 43      | F   | 15 x 10  | Lt. upper outer quadrant  | Surgery      | Germany       | 2006 |
| 18 | Torcasio et al[16]   | 26     | F   | 3        | Rt. inner quadrant        | Surgery      | Italy         | 2006 |
| 19 | Ogun et al[17]       | 38     | F   | 5 x 4    | Lt. upper outer quadrant  | Surgery      | Nigeria       | 2007 |
| 20 | Min et al[18]        | 36     | F   | 4 x 3    | Rt. upper outer quadrant  | Surgery      | Korea         | 2008 |
| 21 | Kwon et al[19]       | 34     | F   | 20 x 17  | Lt. whole breast          | Surgery      | Korea         | 2009 |
| 22 | Sasi et al[20]       | 37     | F   | 10       | Lt. upper outer quadrant  | Surgery      | England       | 2010 |
| 23 | Balaji and Ramachandran[21]| 33      | F   | —        | Lt. whole breast          | —            | —            | 2010 |
| 24 | Malhotra et al[22]   | 60     | M   | 7 x 7 x 5| Lt. upper outer quadrant  | Surgery      | India         | 2010 |
| 25 | Nguyen et al[23]     | 71     | F   | 6        | Lt. axillary              | Surgery      | USA           | 2011 |
| 26 | Gupta and Singh[24]  | 8      | M   | 7 x 6.5 x 3| Rt. upper outer quadrant  | Surgery      | India         | 2011 |
| 27 | Hynes et al[25]      | 33     | F   | —        | Lt. whole breast          | Surgery      | Ireland       | 2012 |
| 28 | Hatbade et al[26]    | 23     | F   | 20 x 10  | Rt. upper outer quadrant  | Surgery      | India         | 2013 |
| 29 | Hirnath and Binu[7]  | 23     | F   | 6 x 7    | Lt. areola lower          | Surgery      | India         | 2014 |
| 30 | Almahdri et al[27]   | 47     | F   | 0.7      | Lt. nipple                | Punch biopsy | USA           | 2014 |
| 31 | Varghese-Hernandez et al[28]| 45      | F   | —        | Lt. whole breast          | Surgery      | Mexico        | 2014 |
| 32 | Almahdri et al[29]   | 37     | F   | 20 x 15 x 10| Lt. upper outer quadrant  | Surgery      | USA           | 2015 |
| 33 | Rusdians et al[30]   | 20     | F   | 3 x 1.5 x 1.5| Lt. inner outer quadrant | Surgery      | USA           | 2016 |
| 34 | Almahdri et al[31]   | 39     | F   | —        | Rt. whole breast          | Surgery      | Saudi Arabia  | 2017 |
| 35 | Chotai et al[32]     | 41     | F   | 3.9 x 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 hemangiomia and lymphangiomia.[24] Lymphatic endothelial markers are CD31, CD34, podoplanin, LYVE-1, and PORX-1. Otherwise, the vascular endothelial marker is VIII-associated antigen.[14] 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.[21] However, these are associated with high rate of recurrence.[17] So complete surgical excision is needed for low risk of recurrence.[10] Spontaneous resolution is uncommon.[21] 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 lymphangiomia should be considered for a prompt diagnosis and definitive treatment to prevent recurrence and complications.[21] Complete surgical excision is the most effective option.

Author contributions

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