

## Case Report

# Cystic lymphangioma of breast and axillary region in an adult: a rare presentation

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**Received:** 18 September 2020

**Accepted:** 06 November 2020

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## ABSTRACT

Cystic lymphangioma also known as cystic hygroma, is a congenital malformation of lymphatic system. Most lymphangioma are present at birth and are diagnosed by the age of 2 years. They are usually located in the head and neck region and are rare in other location. We are reporting a case of cystic lymphangioma in breast and axillary region in a 23 years old female. Physical examination revealed a non-tender cystic mass in axilla and upper outer quadrant of left breast. Ultrasonography (USG) revealed a hypoechoic mass lesion and magnetic resonance imaging (MRI) showed a multi-septated cystic mass in left axilla closely involving the left breast parenchyma. Wide local excision was done and histopathological examination further confirmed the diagnosis of cystic lymphangioma. Although it is very rare, cystic lymphangioma should be considered in the differential diagnosis of mass in breast and axillary region.

**Keywords:** Lymphangioma, Cystic, Breast, Axilla

## INTRODUCTION

A lymphangioma is a malformation of the lymphatic system most commonly observed in infants and children.<sup>1-3</sup> Case reports of lymphangioma in adults are very rare and fewer than 150 cases can be found in the English language scientific literature.<sup>1</sup> Also referred to as cystic hygromas or lymphatic malformations, they are most often found in the cervicofacial region (75%) and are less commonly seen in the axilla (20%) or elsewhere.<sup>1-4</sup> Previous reports have mentioned the management of lymphangiomas in the adult, but these reviews are limited to presentations in the neck.<sup>5,6</sup> A review of literature has identified rare case reports of adult onset lymphangioma of the axilla.<sup>7-14</sup>

The exact nature of lymphangioma formation remains debated. Once considered to be neoplastic in nature, these benign masses are now better understood. Cystic hygroma are present in around one in 2000-4000 live births and 90% are diagnosed before 2 years old.<sup>2,3,16</sup> Some have suggested that trauma or virus infection could be the cause in their

development, while others discuss a more congenital etiology involving miscommunication between lymphatic and venous pathways, aberrant lymphatic growths, and tissue sequestration during development.<sup>2,15</sup> The role of chromosomal abnormalities has been documented, most frequently involving Turner's syndrome, trisomies 13, 18, and 21, and Noonan syndrome.<sup>2</sup> A review of the literature failed to reveal information about the prevalence of chromosomal abnormalities in adult-onset cases, however.

Several staging and classification mechanisms are proposed and adopted which permit for better diagnosis and management. Smith et al. characterized cystic hygromas as either microcystic, macrocystic, or mixed, with microcystic containing cysts <2 cm in diameter. This allowed for a more accurate prediction of how the mass would answer sclerotherapy treatment.<sup>6,17</sup> Mulliken has described lymphangiomas in terms of histological appearance as either capillary lymphangiomas, cavernous lymphangiomas, or cystic hygromas.<sup>18</sup> De Serres proposed a more practical organisation supported location, which allowed for a better estimation of prognosis and surgical

complication rate, but applies only to masses within the cervicofacial region.<sup>19</sup> Diagnosis is often aided by the employment of fine needle aspiration for cytology, tissue histology, and ultrasound, magnetic resonance imaging (MRI) or computed tomography (CT) for definition of the mass.

## CASE REPORT

A 23-year-old, otherwise healthy female was referred to general surgery with complain of swelling in left axilla and breast which was sudden onset and non-tender. She had no history of trauma or surgery to that area. She had no comorbidities, no history of any substance abuse and family history was unremarkable for any congenital masses or chromosomal abnormalities.

On physical examination (Figure 1), she was well-nourished and well developed, with normal temperature, vital signs and normal cardiopulmonary examination. She was found to have a swelling of size around 13×10×5 cm in the left axilla extending till the outer lower compartment of left breast. The swelling was fluctuant, and nontender.

Magnetic resonance imaging (MRI) demonstrated a well-defined T1 hypointense, T2/STIR hyperintense multi-spectated cystic mass in left axilla closely abutting the left breast parenchyma. USG guided fine needle aspiration (FNA) was done and patient underwent surgical excision of the tumour and histopathology confirmed the diagnosis of lymphangioma.

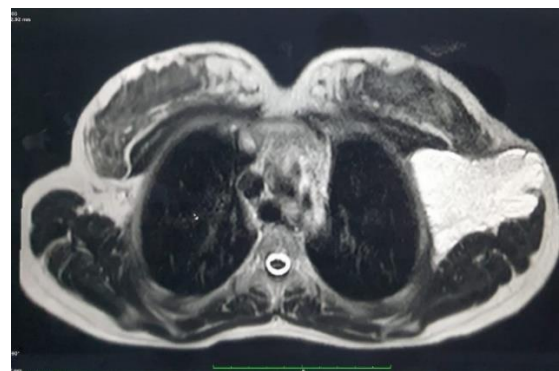


**Figure 1: Mass as demonstrated on physical examination.**

A full excision of the mass was performed under general anaesthesia. The patient was positioned in the left lateral decubitus position and an elliptical incision was given in the axilla. The tumour was dissected away from the pectoralis major and pectoralis minor anteriorly and serratus anterior and subclavius muscle posteriorly. Where the capsule wall was thin and friable suture ligation was done to prevent spillage of contents and to maintain continuity of the structure for its complete removal.



**Figure 2: MRI STIR sequence showing the lesion of size 12.4×9.4×5 cm in left axilla extending till D8 vertebral level.**



**Figure 3: MRI T2 axial view showing lesion involving left breast parenchyma.**

As the dissection continued cephalad, medial pectoral, long thoracic, and thoracodorsal nerves were identified and spared. Where the cyst become embedded in the axilla, gentle blunt dissection was done to remove the remaining mass (Figure 5). The skin was closed in two layers of absorbable sutures and drained with 12 mm closed suction drains.

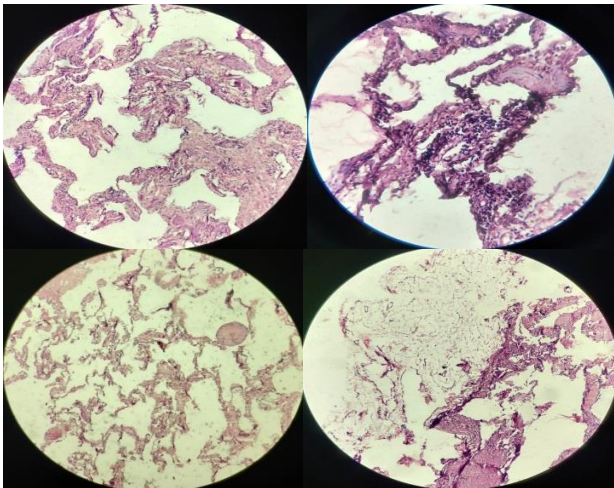


**Figure 4: Intra-operative picture showing tumour freed from adhesions and being taken out.**



**Figure 5: Post-op picture of the excised tumour.**

Cytological analysis of the initial fine needle cytology showed large number of small lymphocytes against mucoid background suggestive of lymphangioma. The full thickness biopsy of the mass showed the presence of loose fibrofatty tissue interspersed with spaces lined by endothelium and at places filled with lymphocytes, polymorphs and red blood cells. The stroma at places showing lymphoid nodule.



**Figure 6: Microscopic section.**

## DISCUSSION

Differential diagnosis of the lymphangioma includes soft tissue sarcoma, abscess, synovial cyst, and hematoma.<sup>18</sup> Surgical excision has been the treatment of lymphangiomas historically, which is believed to be preferred in adults because the lesion is circumscribed.<sup>1-3,5,15</sup> During this case, surgical excision was felt to be particularly uncomplicated with thin encapsulation of the mass, weak adhesions to surrounding tissue, with minimal neurovascular sacrifice. Further, the potential for spontaneous bacterial infection heightens the hazard of delaying therapy in hopes of achieving the spontaneous regression sometimes seen in children.<sup>3</sup> Some concern regarding the surgical therapy exist with the possibility of recurrence. Some reports have mentioned the role of intraoperative cyst rupture making the boundaries of the

mass less clear.<sup>20</sup> However, one study demonstrated that patients having partial resection, just one in nine had recurrence.<sup>21</sup> Several case reports have discussed the inefficacy of a straightforward aspiration and antibiotics.<sup>1,9,13</sup> Long duration follow-up is suggested as a vital aspect of management, as recurrence has occurred as late as 6 years after excision.<sup>15</sup>

Some authors have highlighted the role of sclerotherapy for the management of lymphangiomas, using agents such as OK-432, bleomycin, doxycycline, acetic acid, alcohol, and hypertonic saline.<sup>2,5</sup> sclerotherapy wasn't employed in this case for several reasons given the proximity of the mass to vital neurovascular structures and multicystic anatomy. First, the success of sclerotherapy has occasionally been measured in terms of sufficient mass reduction without complete resolution.<sup>1</sup> Further, it's been recognized that sclerotherapy might not be effective against multiloculated masses, or those of mixed or microcystic anatomy.<sup>1,2,5</sup> Sclerotherapy may induce a localized immune response which causes a short lived but dramatic increase within the size of the mass. Perkins et al has also recognized the potential for shock-like reaction in the setting of penicillin allergy and OK-432 use.<sup>5</sup>

## CONCLUSION

Continued reporting of adult-onset cystic lymphangioma is of particular importance, as the nature and management of these rare masses are elucidated. The role of chromosomal abnormalities in cases presenting in the adult is not yet understood and may be of interest as such masses continue to be reported. This case contributes to the body of evidence supporting the role of cystic lymphangioma in a differential diagnosis for masses in the adult, especially in the acute setting. Further, the future management of cystic hygromas in the axilla, with proximity to important neurovascular structures, is better informed with our addition to examples of uncomplicated resection.

*Funding: No funding sources*

*Conflict of interest: None declared*

*Ethical approval: Not required*

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**Cite this article as:** Shukla S, Shivhare R, Lal V, Rathore D. Cystic lymphangioma of breast and axillary region in an adult: a rare presentation. *Int Surg J* 2021;8:391-4.