

Cystic Lymphangioma of the Breast in an Adult Woman - A Case Report

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Abstract: *Lymphangiomas are congenital malformations of lymphatic vessels. More than 50% of these lesions are present at birth; 90% are diagnosed by the age of 2. These lesions do not expand very rapidly but they tend to infiltrate surrounding tissues; their degeneration into malignant tumors is an extremely rare occurrence. They are mostly located in the neck region and the axilla; breast lymphangioma is very rare. Surgery is usually performed for aesthetic reasons and in order to make a differential diagnosis with other, more common lesions. The surgical procedure involves the excision of the mass; other methods, such as radiotherapy and sclerotherapy, have proved to be completely ineffective. Although it is very rare, cystic lymphangioma should be considered in the differential diagnosis of a breast mass in adults*

1. Introduction

Cystic lymphangioma, also known as cystic hygroma, is a rare benign congenital malformation of the lymphatic system [1]. It is a vascular malformation rather than a true neoplasm, and this lesion is believed to be the result from failure of the lymphatic system to connect with the venous system. Possible etiological factors include Blockage of the lymphatic channels with resultant secondary dilatation, congenital weakness of the lymphatic wall or proliferation of lymphatic vessels [2-4]. It may be associated with other anatomic and/ or chromosomal abnormalities. More than 90% of cystic hygroma cases are diagnosed by the age of 2 years, in more than 90% of cases the lesion is located in the neck or axilla [1,5].

It is a rare condition to occur in the breast and only few cases have been reported. Therefore, we report an additional case with a typical clinical and radiological appearance, confirmed by histopathological analysis. Here in this article, we report a case of cystic lymphangioma of breast in a 26-year old woman with diagnosis and treatment.

2. Case Report

A 26 yr old female patient with no known co morbidities presented to the OPD with a lump in the right breast from past 7-8 months which was insidious onset and gradually progressed in size to the present size.

Patient does not give history of pain, nipple discharge, axillary swelling, breathing difficulty, back pain or other bony pains etc.

No significant family history

Her general examination was normal and on local examination of the left breast and left axilla was normal. Examination of the right breast revealed a solitary lump measuring 25*15 cms in its greatest dimensions, tender on touch, present in right upper outer quadrant, soft in consistency. Nipple areola complex was normal with no palpable axillary nodes.

Rest of breast examination was unremarkable.

Routine laboratory blood investigations were normal.

MRI of the right breast:- lobulated multiseptated cystic lesion with intermuscular extension suggestive of cystic lymphangioma of right breast.

3. Course in the Hospital

Preoperatively pre anaesthetic clearance was obtained and patient consented for proposed surgery after understanding all the possible complications associated with the surgery.

Intraoperatively multiple fluid filled, poorly demarcated cystic lesions containing clear yellow fluid was noted and entire mass was removed in toto.

Postoperative period recovery was uneventful and suture removal done on the 7th day after complete wound healing.

Histopathology report showed dilated lymphatic spaces of different size lined by flat endothelium and patient was lost on further follow up.



Figure 1: Large Cystic Lesion Measuring About 25*15 Cms in the Upper Outer Quadrant of the Right Breast

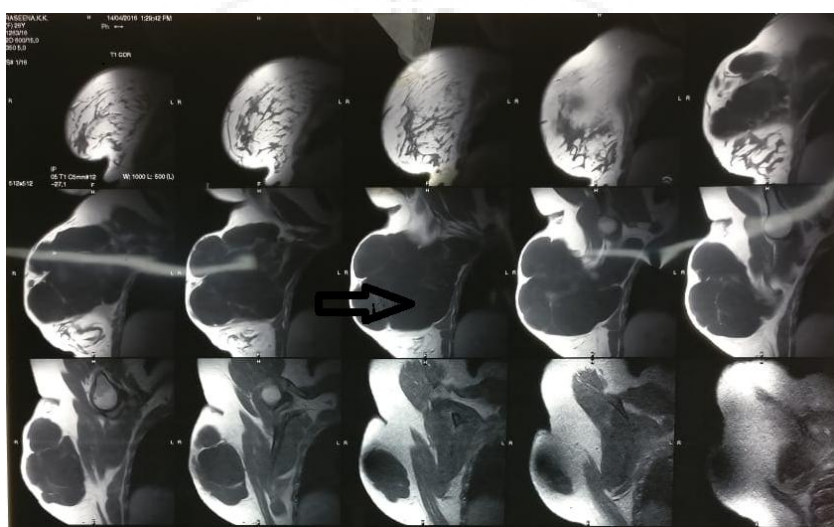


Figure 2: MRI of the Right Breast Showing Lobulated Multiseptated Cystic Lesion with Intermuscular Extension Suggestive of Cystic Lymphangioma of the Right Breast

Arrow Pointing Towards Cystic Lymphangioma

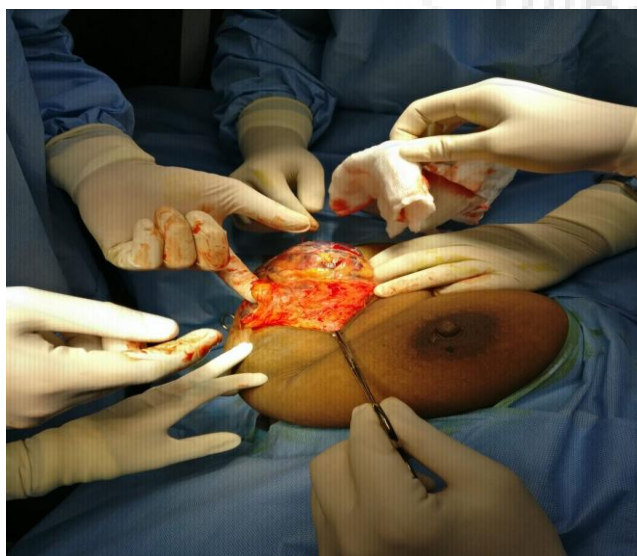


Figure 3: Intraoperatively Multiple , Poorly Demarcated , Cystic Lesion Containing Clear Yellow Fluid

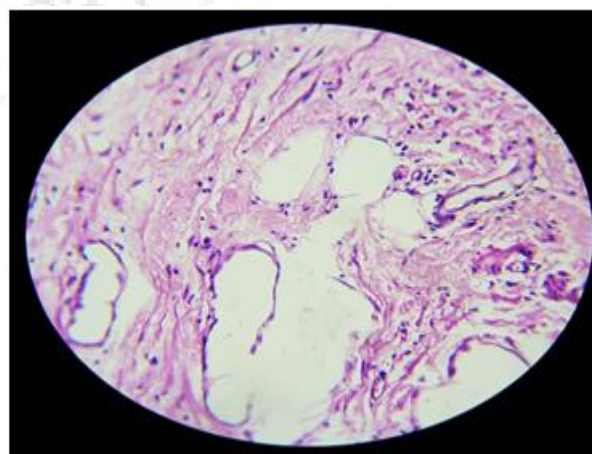


Figure 4: Histopathologically shows Dilated Lymphatic Spaces, of Different Size, Lined by Flat Endothelium

4. Discussion

Cystic lymphangioma is a malformation that arises from sequestration of lymphatic tissue that fails to communicate in a normal fashion with the lymphatic system, that is believed to result from a failure of the lymphatic system to connect with the venous system [4-6]. The other etiologic factors of cystic lymphangioma include obstruction of the lymphatic channels with secondary dilatation, congenital weakness of the lymphatic wall and proliferation of the lymphatic vessels [7]. 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 [1-3].

Lymphangioma can be diffuse or multi-centric. They can be located in the soft tissues only, or can extend to involve other locations like the bone and the viscera. Around 70% of lymphangiomas occur in the neck region, 20% in the axilla, while the remaining 10% is reported in the abdominal organs, retro peritoneum, skeleton, scrotum and, very rarely, in the breast [1,4].

Lymphangiomas at ultrasonography usually appear as a cystic multiloculated mass, with linear septa of variable thickness that contain solid elements originating from the cyst walls or septa. Larger lesions will not have well defined margins and cysts may extend to surrounding tissues [8,9]. The mammographic findings are non-specific as many of the cases occur high in the axilla, and its visualization on mammogram might be difficult. Magnetic Resonance Imaging (MRI) is proposed to be the best modality for the diagnosis and evaluation of lymphangioma extension [10]. Cystic lymphangioma appears as multiseptated masses on MRI, usually following the signal intensity of fluid, showing low signal intensity on T1 and high signal intensity on T2, with enhancement of the septa. The differential diagnosis should include simple cysts, postsurgical liquid collection, hematoma, lymphocele, abscess and hemangioma.

Primary treatment of lymphangioma is complete surgical excision. Ensuring safe margins is important to prevent recurrence. The greatest difficulty in treatment is related to the size and location of the mass as it may be difficult to obtain safe margins due to the tendency of these lesions to infiltrate surrounding tissues and hence there is a high risk of incomplete excision, possibly leading to recurrence [6-8]. Other options are sclerotherapy, incision and drainage, irradiation, and cryotherapy which have proved to be ineffective and may carry risks such as hemorrhage and infection [10].

5. Conclusion

Cystic lymphangioma, is a congenital malformation of the lymphatic system. Most lymphangiomas are present at birth and they are diagnosed in the early age. The breast as a site of origin, is an extremely unusual location, and especially in adults. Although the incidence of this disease is extremely rare in the adult breast, cystic lymphangioma should be considered in the differential diagnosis of an irregular cystic

mass of the breast. Complete surgical excision is the treatment of choice.

6. Consent

Informed written consent was obtained from the patient based on whom the report is about.

7. Conflict of interest

The authors declare that they have no conflict of interest.

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