CYSTIC LYMPHANGIOMA IN AN ADULT-DIAGNOSIS ON CYTOLOGY

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PRESENTATION OF CASE

A 63- year old lady presented with a large mass in the right side of the chest since 1 year, which was gradually increasing in size. The swelling was associated with pain for the last 6 months. The patient did not give any history of trauma. On examination, the mass was seen extending from the upper outer quadrant of the right breast medially, up to the right axilla laterally. The swelling measured 10 x 10 cms, and was soft, fluctuant, tender, possibly cystic [Figure 1].

Clinically diagnosed as lipoma and radiologically as cystic disease of the breast and abscess, the diagnosis of lymphangioma was rightly suggested by fine needle aspiration cytology and confirmed with histopathology. This case highlights the role of FNAC in the diagnosis of these lesions. Presence of cystic hygroma in adults makes the present case report unique and interesting.

Lymphangiomas or cystic hygromas are cystic lesions commonly seen in the soft tissues of the neck, usually in the paediatric age group. Axilla and breast are rare sites, more so in adults. Lymphangioma is a benign lesion, composed of dilated lymphatic channels.¹ They are common paediatric lesions and are thought to represent developmental malformations rather than true neoplasms.^{1,2} Lymphangiomas are relatively rare; more so in adults. They can arise anywhere in the body, with a predilection for the head, neck, and axilla. Their occurrence in the breast is particularly rare and few cases have been reported in literature.^{3,4}

CLINICAL DIAGNOSIS

With the above examination findings, differential diagnoses of lipoma or cystic disease of the breast or chest wall were considered.

Figure 1. Soft swelling extending from the upper outer quadrant of the right breast to the axilla

Further investigations were ordered, including mammogram and ultrasonogram [USG]. The findings were as follows:

Mammogram showed a well-defined radio-opaque lesion, with irregular margins, in the superolateral aspect of right breast, suggestive of benign lesion/ BIRADS 2.

On ultrasonogram, a large, well- defined round to oval hypoechoic lesion was described in the right breast, measuring 6.8 x 5.7 x 5.6 cms, with internal septations and internal echoes in the intramuscular plane. The mass did not show increased vascularity. Breast parenchyma was normal. No axillary lymph nodes or focal lesions were reported. With these findings, a sonologic diagnosis of intramuscular abscess, with internal septations and echoes, was offered [Figure 2].

Figure 2. Hypoechoic lesion measuring 6.8x5.7x5.6 cms, with internal septations and internal echoes in the intramuscular plane

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With the above radiologic opinions, further clarification was sought with cytology and FNAC was requested.

Fine Needle Aspiration Cytology

Using a Cameco gun, 10 ml syringe and 22- gauge needle, about 20 ml of straw coloured clear fluid was aspirated [Figure 3]. The swelling reduced in size considerably.

Figure 3. Straw coloured clear fluid aspirated from the swelling

Both air-dried and alcohol-fixed smears were prepared from the centrifuged deposits. Air-dried smears were stained with Giemsa stain and alcohol-fixed smears were stained with Haematoxylin and Eosin stains.

On microscopy, direct and sediment smears showed plenty of only mature small lymphocytes and occasional macrophages, in a proteinaceous background. No epithelial cells or atypia were noted. With these findings, the swelling was reported as a benign cystic lesion, with the possibility of cystic lymphangioma to be considered [Figure 4].

Figure 4. Sediment smears showing mature lymphocytes in a proteinaceous background [20x; H&E on left and Giemsa on right]

The cyst was excised and submitted for histopathology.

Histopathologic Examination

Gross: an irregular soft tissue mass was received, measuring 9x4x2.5 cms, with multiple cystic spaces on cut surface [Figure 5].

Figure 5. Collapsed cysts on the cut surface of the excised mass

Microscopy

Sections showed large lymphatic channels lined by flattened epithelium. Many of the spaces were empty and some were filled with lymph and few erythrocytes. They were surrounded by fibrocollagenous stroma, collections of lymphocytes, dilated blood vessels, muscle bundles and adipose tissue. These features confirmed a diagnosis of cystic lymphangioma [Figure 6].

Figure 6. Photomicrographs show lymphatic channels, some empty, some filled with lymph and RBCs. Lymphocyte aggregates, blood vessels, disorganised muscle bundles and adipose tissue is seen in the surrounding stroma. [H&E, 4x]

DISCUSSION

Lymphangioma is defined as a benign, cavernous/ cystic vascular lesion, composed of dilated lymphatic channels.¹ They may exist in three forms: capillary, cavernous and cystic.² Traditionally called hygromas, cystic lymphangiomas are common paediatric lesions, most often presenting at birth or during the first years of life. Up to 90% of cases present by the second year of age.⁵

Lymphangiomas are relatively rare, and uncommon in adults. They can arise anywhere in the body, with a predilection for the head, neck, and axilla. They occur sporadically in the lung, gastrointestinal tract, spleen, liver, and bone.³ Their occurrence in the breast is particularly rare and very few cases have been reported in literature. The upper outer quadrant of breast and axillary tail of Spence, the reported sites of breast lymphangiomas, are related anatomically to the drainage route of the breast lymphatics, towards the tail and axilla.^{5,6}

Congenital or early appearance in life and architecture of the lesion favours developmental malformations as an etiologic factor rather than true neoplasms, with genetic abnormalities playing an additional role. They possibly arise

as sequestrations of lymphatics that fail to communicate with the venous system.^{1,2} They may also have some capacity to proliferate.³

Histological classification of lymphangiomas into subtypes based on vessel size is no longer used. Many have both cavernous and cystic components, suggesting the possibility of a long standing cavernous evolving into a cystic type. This is possibly attributed to their anatomic location. Cystic lymphangiomas arise in the neck and axilla, where loose connective tissue allows expansion of these channels; cavernous type develops in the mouth, lips, cheek, tongue, or other areas where dense connective tissue and muscle prevent expansion.³

The lesions present as circumscribed painless swellings and are soft and fluctuant at palpation.¹ They present in children as a poorly defined soft tissue mass in the neck.² They may enlarge, remain static, or wax and wane during the period of clinical observation.³ They may rapidly expand due to haemorrhage, infection or trauma. The cystic swelling is brilliantly transilluminant clinically. It gets recurrently infected because of its lymphoid content.⁴

They vary greatly in size, ranging from 3 to 25 cms in diameter.⁵ Ultrasonography display their cystic nature, angiography shows poor vascularisation and CT scan reveals multiple, homogeneous, non- enhancing areas.¹

DIFFERENTIAL DIAGNOSES

Include cystic diseases of the breast, abscess, lymphangiectasis, lesions of the lymph node and soft tissue. Sonography though helpful, may not always give a definitive diagnosis. Fine needle aspiration cytology is most useful in these cases.⁵

Cytology

There is no consensus in the literature regarding the use of FNAC to diagnose these lesions.⁷ It only allows a suggested diagnosis. According to Orell et al,⁸ smears of fluid aspirated from a cystic lymphangioma contain only lymphoid cells, mainly small lymphocytes, in variable numbers.

In a study of 20 cases of cystic lymphangioma by Alka et al,⁴ the most common site was the neck (90.0%), followed by mediastinum (5.0%) and axilla (5.0%- 1 case). Of the 20 cases radiologically diagnosed as "possibly cystic hygroma", 15 were confidently called so on FNAC. Lymphoid cells [in 17 aspirates], endothelial cells [in 10 aspirates], cholesterol crystals [in 10 aspirates] and red blood cells [in 2 aspirates] were reported on cytology. Background had proteinaceous material in 15 aspirates.

FNAC is a rapid, convenient and accurate diagnostic tool, that can be done on an outpatient basis. The procedure is safe and free from complications and is well tolerated by patients. Along with radiological correlation, it serves as a highly effective and efficient modality for the confident diagnosis of cystic hygroma especially in patients with atypical presentation, age and location.⁴

Histopathology

Grossly, cavernous/cystic lymphangiomas appear as multicystic or spongy mass, containing watery or milky fluid in the cavities.¹ They vary from well-circumscribed lesions with interconnecting cysts to ill-defined, sponge-like compressible lesions composed of microscopic cysts.³ On

light microscopy, they are characterised by large, thin walled, dilated lymphatic vessels of different size. The small lymphatics resemble capillaries, and sometimes tentatively identified from their contents. The spaces are lined by attenuated endothelium resembling normal lymphatics. Unlike capillaries, it is neither invested by basement membrane nor pericytes. Larger spaces can be invested by fascicles of poorly developed and disorganised smooth muscle. The lymphatic spaces classically are filled with proteinaceous fluid containing lymphocytes, although occasionally erythrocytes are present as well. The stroma is composed of collagen meshwork, punctuated by small lymphoid aggregates. Long standing lesions reveal interstitial fibrosis and stromal inflammation. Histologic features that favour the diagnosis of lymphangioma include the presence of lymphoid aggregates in the stroma and irregular lumens with widely spaced nuclei. Immunohistochemistry for lymphatic lineage markers (e.g., VEGFR3, D2-40) can help in this distinction.^{1,2,3}

FINAL DIAGNOSIS

Based on the cytology findings, a diagnosis of cystic lymphangioma was suggested in the present case and the same was confirmed with histopathology. Although benign breast disease and abscess were considered on radiology, the two were ruled out following aspiration of large volumes of clear fluid, which contained only mature lymphocytes on cytology and owing to lack of epithelial cells, necrosis or neutrophilic infiltrate either on cytology or in tissue sections.

Treatment

Surgery is the treatment of choice.⁶ Lymphangiomas are benign lesions, and therapy is dictated largely by their location and clinical extent.³ Tendency of these lesions to infiltrate the surrounding tissues makes it difficult to obtain clear margins, in which case they tend to recur.⁶

CONCLUSION

Cystic lymphangioma is uncommonly seen in clinical practice. Unusual presentations and sparing literature add to their diagnostic complexity. Lymphangioma in the axilla and breast is rare, and should be borne in mind, while investigating cystic lesions in these locations. Cytology is a useful investigative modality and FNAC is one of the diagnostic techniques often requested in such cases. It can be diagnostic when the lesion is harboured among the differential diagnoses. Diagnostic efficacy can further be improved when combined with imaging techniques like ultrasound and computerized tomography scans.

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