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Original Research Paper

Pathology

CYTOPATHOLOGY AS AN IMPORTANT TOOL IN DIAGNOSING LYMPHANGIOMA CIRCUMSCRIPTUM OF BREAST – A RARE CASE REPORT.

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ABSTRACT Lymphangiomas of breast are rare with only a few cases being reported so far. Among these secondary lymphangioma have been reported but cases of primary lymphangioma are even rarer. Due to its proclivity to mimic malignancy clinically, its accurate diagnosis plays a crucial role in management of these patients. This is a unique case where cytology suggested a diagnosis of vascular malformation ruling out malignancy, thus avoiding unnecessary surgical exploration along with untoward effects of chemo/radiotherapy. The histopathology revealed diagnosis of lymphangioma circumscriptum of breast.

KEYWORDS : Lymphangioma circumscriptum, rare, breast.

INTRODUCTION:

Lymphangiomas, now considered as lymphatic malformations, are regarded as abnormality of morphogenesis rather than as a neoplasm. Cutaneous lymphatic malformations are divided into superficial and deep forms, lymphangioma circumscriptum (LC) being superficial type. The most common site for LC is limb and limb girdle with few cases of secondary LC being reported in breast following mastectomy and radiotherapy. [1] However primary LC of breast is extremely rare with exact incidence not known. Hence we report this unique case having clinical suspicion of malignancy.

Case History: A female of 30 years came to surgery OPD with superficial plaque- like reddish brown cutaneous lesions showing scaly patches and multiple tiny vesicles in lower outer quadrant of right breast. (Figure 1) The lesion started after a trivial trauma 15 years back which began as a small reddish tiny plaque progressing gradually to the current size. There was no associated nipple discharge or lump or any other clinical symptoms except for itching and scanty intermittent bloody discharge at the site of lesion. On examination, right breast showed similar lesions as described above confined to the skin only and involving an area of 5x5 cm. The adjacent skin showed a rim of multiple tiny reddish spots. Underlying breast parenchyma did not reveal any obvious lump. Radiological examination suggested differential diagnosis of angiosarcoma and malignant melanoma. With these differentials the patient was referred to cytology and FNAC was performed. The aspirate was blood mixed, the smears of which were prepared and stained with Leishman and Hematoxylin and Eosin stains (H. & E.). The microscopy revealed multiple vascular channels with scanty endothelial cells against few lymphoid cells and red blood cells in background which favoured a diagnosis of vascular malformation. (Figure 2A &B) Following this the patient underwent wide local excision of the mass and the specimen was sent for histopathological examination. The gross examination revealed a skin covered specimen of size 13x8x2 cm with the overlying skin showing the lesion as described above involving an area of 5x5 cm with tiny grey black maculonodular lesions in the surrounding skin. (Figure 3A) However the resection margins were apparently unremarkable. On serial sectioning, the lesion was limited to the dermis with no involvement of subcutaneous tissue. Multiple sections were taken, stained with H. & E. and studied. The microscopy revealed multiple dilated lymphatic channels in the superficial and papillary dermis containing clear pale eosinophilic fluid admixed with lymphocytes. (Figure 3B)Thus final diagnosis was given as superficial capillary lymphangioma (lymphangioma circumscriptum) of breast.

DISCUSSION:

Lymphangiomas are benign lymphatic tumours which are developmental in origin and so are common in children, with 90% of the cases presenting in the second decade of life. [2]They are now considered as lymphatic malformations and regarded as abnormality of morphogenesis rather than as a neoplasm. They are sub classified as microcystic, macrocystic (cyst greater than 0.5 cm) or combined.[3] Diffuse lymphatic malformation involving multiple organs are still often referred to as lymphangiomatosis.[4] Cutaneous lymphatic malformations are divided into superficial and deep forms, lymphangioma circumscriptum (LC) being superficial type. The first reported description of LC was done by Fox and Fox in 1878 who named it as lymphangiectodes. [5, 1]The term LC was first used by Morris in 1889.[6,1]The most common sites of LC are mucosa of mouth, tongue, groin, axilla, trunk and proximal region of extremities.[7] However, LC of breast is rare with few case reports of secondary LC being reported following mastectomy and radiotherapy. However primary LC of breast is even rarer with very few cases reported.

The etiology is not fully understood, but the majority of cases are thought to represent congenital abnormality, possibly resulting from sequestered island of lymphatic tissue that develops without any connection to normal lymphatic channel. Trauma including surgery and radiation therapy has been implicated in some cases as the causative agent. The clinical presentation includes multiple lesions which are generally localized to one specific area. When they are confined to an area less than 7 cm in diameter they are referred to as LC while those greater than 7 cm are referred to as lymphangioma diffusum.[1]On microscopy they reveal multiple dilated lymphatic channels involving the superficial and papillary dermis. The histology features that favours

lymphatic malformation over vascular malformation are multiple empty or spaces filled with proteinaceous fluid containing lymphocytes, more irregular lumens with widely spaced nuclei, inconspicuous media and adventitial coat and lymphoid aggregates in the stroma. Thus LC can be easily diagnosed based on correlation of clinical morphology and histology making immunohistochemistry (IHC) a secondary tool for further confirmation. The IHC markers for lymphatic lineage are PROX1, VEGF3 and D2-40 (podoplanin), first two showing greater sensitivity.[8]

The cytology findings of LC have the least mention in existing literature with no specific criteria mentioned. However, suggestive findings include multiple thin vascular channels lined by scanty endothelial cells seen against a fluidy background containing scanty lymphocytes. As observed in this case, the uneven and widely spaced arrangement of endothelial cells favour lymphatic malformation over vascular in which they are even and regularly spaced.

As primary LC being an extremely rare entity described in breast and clinically mimics angiosarcoma or melanoma, it leads to misdiagnosis. The natural course of these lesions is slow progression of the lesion both in size and area with leakage of fluid, infection, local discomfort from hypertrophy and irritation from clothing and movement. Although it shows a benign course, it can cause significant morbidity due to its large size, location or proclivity to become secondarily infected. Only rare cases are known to regress spontaneously. Therefore mainstay of therapy is surgery.The excision includes deep lymphatic cisterns with wide subcutaneous tissue followed by primary skin closure or skin grafting. The other modality of treatment available includes radiotherapy, electro cautery, carbon dioxide laser therapy and topical dressings.

In this case the cytology findings suggested the diagnosis of vascular malformation following which patient underwent wide local excision, which on histopathology was confirmed as LC. Thus the cytology ruled out malignancy and avoided unnecessary exploration and radio/chemotherapy. Hence cytology can be used as an important diagnostic tool in streamlining the management with additional benefits of it being rapid and inexpensive.

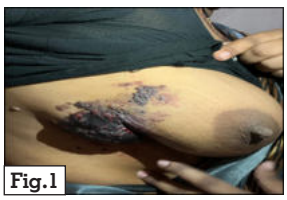
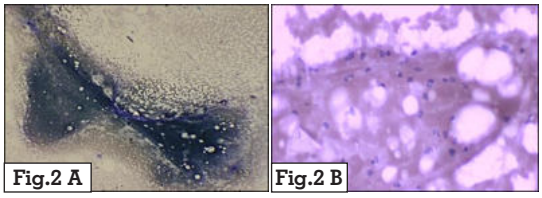
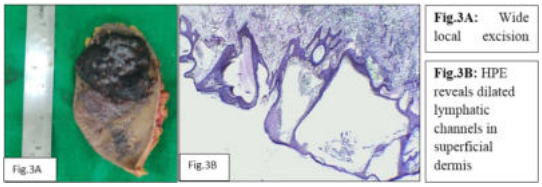


Fig. 1: Photograph showing lesion in right breast upper-outer quadrant.
Fig. 2A: Multiple vascular channels.(H and E x 100).
Fig. 2B: Endothelial lining seen as scanty spindle cells. (H and E x 400).





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