

Varied presentations of ectopic breast – polymastia, fibroadenoma, and carcinoma arising from ectopic breast tissue

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ABSTRACT

Ectopic breast is a congenital anomaly of the breast which can have varied presentations because of its different sites and pathologies arising from it. Lesions of ectopic breast tissue (EBT) are commonly seen due to persistence of embryonic remnants along the milk line. They have also been reported from other sites like face, vulva, and perineum. They are prone to the same physiological and pathological alterations seen in the normal breast. Only 0.3% of breast carcinomas arise in the ectopic breast, whereas only a few cases of fibroadenoma have been reported at this site. We present a case of polymastia in a 21-year-old female in the inframammary region. We report two cases of fibroadenoma and carcinoma arising from EBT in the axilla of a 26 and 45-year females. Fibroadenoma was treated by simple excision while for carcinoma modified radical mastectomy was done followed by radiation and chemotherapy. Patient developed metastasis in the sternum. Carcinoma arising from EBT has a poorer prognosis and needs early diagnosis and treatment.

Key words: Axilla, carcinoma, ectopic breast, fibroadenoma

INTRODUCTION

During 5th or 6th week of embryonic development, the mammary ridges (milk lines) develop and run from the anterior axillary folds to the groin which usually regress elsewhere except in the pectoral region where it forms the normal breasts. Failure of regression of mammary ridges in other areas apart from the pectoral region is termed as ectopic breast tissue (EBT) seen in about 0.4–6% women.^[1] The most common site is axilla, accounting for approximately 55–65% of EBT.^[2] EBT is prone to all types of benign and malignant pathologies as naturally positioned breasts. However, pathologies of EBT are rare, carcinoma being most frequent, followed by mastopathy and fibroadenoma.^[3] Diagnosis of EBT is significant as it is associated with anomalies of urinary and cardiovascular

system.^[1] Supernumerary breast in the inframammary region, fibroadenoma, and invasive ductal carcinoma arising from EBT in the axilla of ectopic breast are being reported to highlight the varied presentations of EBT.

CASE REPORTS

Case 1

A 21-year-old female presented with the swelling below her left breast since 3 years. On examination, swelling had an underdeveloped nipple and areola, which patient mentioned to be present since birth. Swelling measured 7 cm × 3 cm and was soft and nontender [Figure 1]. Both the breasts and axillae were normal. Ultrasonography of abdomen showed no abnormality. Fine needle aspiration cytology (FNAC) revealed few fragments of fibroadipose tissue with occasional cluster of benign ductal cells. So a diagnosis of supernumerary breast was made. As the patient was not willing for surgery, she had been advised regular follow-up.

Case 2

A 26-year-old female presented with a mass in the right axilla since 2 years which was gradually increasing in size. On examination, 4 cm × 3 cm size swelling was noted in the

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right axilla [Figure 2a]. It was firm in consistency, nontender, freely mobile, and completely separate from the right breast. Skin over the swelling was normal, with no nipple or areola made out. A differential diagnosis of lymphadenopathy or adnexal lesion was made. Ultrasonography of the right axilla suggested the presence of a lymph node [Figure 2b]. A diagnosis of fibroadenoma was made on FNAC, which was confirmed on excision [Figure 3a and b].

Case 3

A 45-year-old premenopausal, multiparous female complained of rapidly progressive left axillary lump since 2 months. On examination, 4 cm × 3 cm lump was present in the axilla. Lump was firm to hard to feel and was fixed to both skin and underlying tissue. On sonomammography, left axillary fold showed a large hypoechoic lesion measuring 3.7 cm × 1.6 cm × 1.6 cm with ill-defined margins. The continuity with the adjacent normal breast parenchyma was not established. Furthermore, there were two irregularly outlined hypoechoic lesions measuring 2.8 cm × 2.0 cm and

2.7 cm × 2.2 cm noted near the main lesion in left axilla. An impression of space occupying lesion in the EBT with axillary lymphadenopathy (BIRAD-5) was given [Figure 4a]. Both breasts were normal. FNAC confirmed the diagnosis of malignancy. Patient underwent left modified radical mastectomy with axillary clearance. Grossly serial cut section from axillary tissue revealed tumor mass measuring 3.5 cm × 3 cm × 2 cm. Grossly axillary lymph nodes appeared metastatic. On microscopy impression of infiltrating duct carcinoma (not otherwise specified) grade III was given [Figure 4b]. 4 out of 14 lymph nodes showed metastasis with the perinodal extension. On immunohistochemistry, tumor was estrogen receptor/progesterone receptor-positive and cerbB2 was equivocal. Bone scan revealed increased uptake with lytic sclerotic component in the sternum. Patient was given 8 cycles of chemotherapy (cyclophosphamide, doxorubicin, and 5-fluorouracil) with tamoxifen and 8 Gy single fraction radiations over sternum. She is on follow-up and has been taking tamoxifen.



Figure 1: Clinical photograph of supernumerary breast with ill-defined nipple and areola

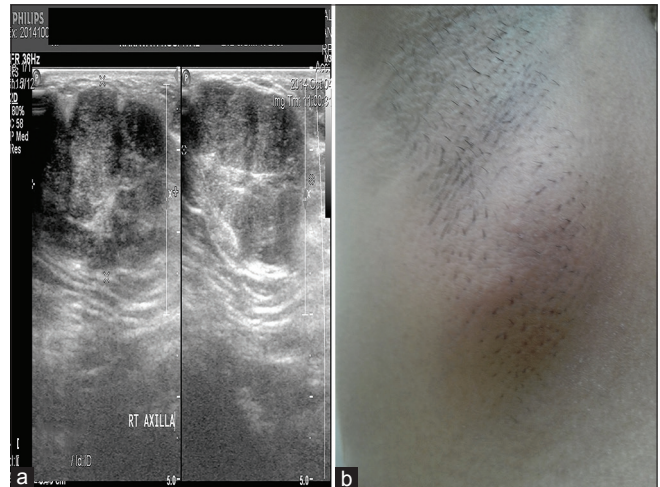


Figure 2: (a) Clinical photograph showing lump in the right axilla. (b) Ultrasound of right axilla showing heterogenous mass lesion

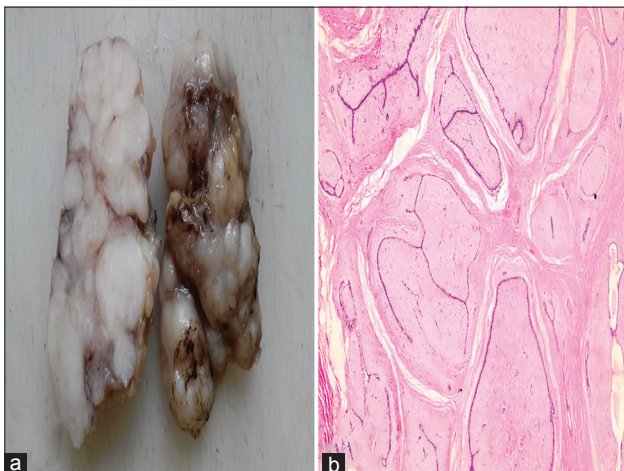


Figure 3: (a) Gross photograph showing well circumscribed white nodule (b) Microscopic photograph showing duct and stroma of fibroadenoma

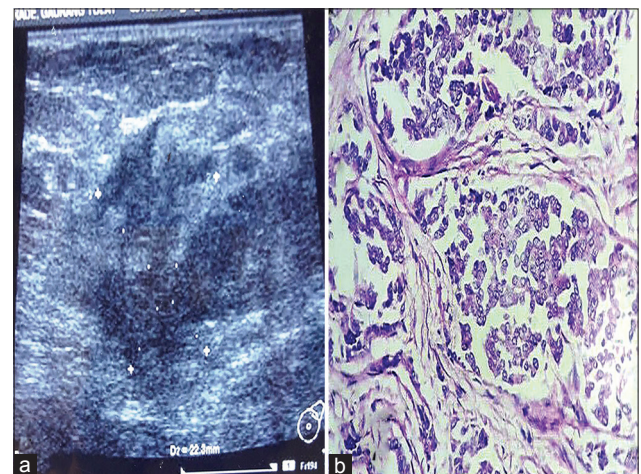


Figure 4: (a) Mammographic photograph showing an irregular hypoechoic mass lesion in the left axillary fold with surrounding fatty breast parenchyma. (b) Microscopic photograph showing infiltrating duct carcinoma

DISCUSSION

Incomplete involution of mammary ridges in the extrapectoral region results in the development of EBT. It may contain all three elements—parenchyma, nipple, and areola or any combination of them and is hormone responsive.^[4] EBT was initially classified by Kajava in 1915 into eight classes.^[5]

Class I consists of a complete breast with nipple, areola, and glandular tissue. Class II consists of the nipple and glandular tissue but no areola. Class III consists of the areola and glandular tissue but no nipple. Class IV consists of glandular tissue only. Class V consists of nipple and areola but no glandular tissue (pseudomamma). Class VI consists of a nipple only (polythelia). Class VII consists of an areola only (polythelia areolaris). Class VIII consists of a patch of hair only (polythelia pilosa).^[5] The present case 1 belongs to class I and case 2 and case 3 fall in class IV category.

In recent literature, EBT is classified as supernumerary breast and an aberrant breast.^[6] Supernumerary breast has nipple, areola or both in combination with ductal tissue. They are usually present along the embryonic milk line, but can occur at other sites like cheek, neck, shoulder, midline of chest or abdomen, flank, hip, thigh, and buttock. On histology, they have an organized duct system communicating with the overlying skin. They also have been associated with supernumerary kidneys, renal agenesis, renal malignancies, pyloric stenosis, epilepsy, and cardiac abnormalities due to corresponding development of mammary tissue and the genitourinary system.^[6] While aberrant breast is an island of breast tissue without a ductal system or relationship to the overlying skin (without nipple or areola) and are usually located in close proximity to the normal breast. Axilla is the most common location, however infraclavicular, juxtasternal, subscapular, epigastric, and vulvar locations have also been described and they are unassociated with other anomalies.^[6] Incidence of EBT is around 0.4–6% and pathologies of EBT are even rarer.^[1,2] Incidence of ectopic breast carcinoma ranges from 0.3–6%, while only few cases of fibroadenoma of the ectopic breast has been mentioned in the literature.^[1,6]

Ectopic breast tissue usually stay dormant until puberty, pregnancy or lactation and are often noticed due to hormonal stimulation causing psychological and cosmetic impact as in our first case.^[7] However, clinical significance lies in the fact that it is at a risk of developing breast diseases such as inflammation, fibrosis, cyst, papillomas, fibroadenoma, duct hyperplasia, cystosarcoma phyllodes, and carcinoma.^[8,9]

Axillary EBT has a broad range of clinical differential diagnosis from benign to malignant lesions such as lipoma,

lymphadenitis, lymphoma, metastatic lymphadenopathy, sebaceous cyst, suppurative hidradenitis, adnexal lesions, and axillary tail of Spence which is more deeply located.^[8,9]

Diagnosis of pathologies of EBT is often delayed and ignored due to its low incidence. Sonomammography helps in distinguishing EBT from the axillary tail of Spence, in defining the nature of the lump and in excluding or detecting associated breast pathologies. However, FNAC and tru-cut biopsy confirms the diagnosis.^[6,8] For asymptomatic EBT, no treatment is indicated though some authors recommend its surgical removal to prevent any subsequent complications or for cosmetic reasons. Liposuction is also a new feasible alternative in selected cases.^[6,8] Treatment for fibroadenoma is simple resection. Treatment protocol for ectopic invasive carcinoma includes a wide local excision with lymph node clearance.^[6] However, Marshall *et al.* noted in his large series of 82 cases, that three cases subsequently developed ipsilateral pectoral breast carcinoma and hence advocated prophylactic ipsilateral mastectomy.^[10] While Evans and Guyton study stated no additional advantage of radical mastectomy over limited axillary mastectomy with lymph node dissection.^[11] Several authors have stressed upon the poor prognosis of carcinoma of EBT owing to early lymph node metastasis, delayed diagnosis, and difficulty in achieving enblock resection of the tumor.^[11,12]

CONCLUSION

Although not a frequent finding, EBT must be kept in mind while dealing with the swellings in the axillary region. Careful investigation must be emphasized as EBT is affected by both benign and malignant diseases. A simple FNAC can help to differentiate EBT from other lesions of axilla.

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