A rare cancer in an unexpected ectopic

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Abstract---Accessory Breasts can occur anywhere along the milk line extending from the axilla to the groin. In view of cosmetic reasons, accessory breasts are removed. We present three cases of accessory breasts with intra-cystic papillary carcinoma in them. Intra-cystic papillary carcinoma is a rare carcinoma that has finger like projections. It has a good prognosis and hence a simple excision biopsy is enough for these patients.

Keywords---rare cancer, unexpected ectopic, accessory breasts.

Introduction

Accessory breasts are defined as extra and / or ectopic breast tissue. They occur due to the failure of the embryonic mammary ridge to regress. They can occur anywhere along the milk line (Axilla to the groin) The commonest site is anterior axillary line and less common sites include axilla, scapula, thigh and labia majora. (1) They also undergo a similar disease processes as normal breast tissue. They can occur with or without the nipple and areola. The occurrence of accessory breast is about 0·4–6 per cent of the general population. (2) Of these one third are bilateral and more common among Asian women. (3) Ethnic dresses worn by Indian women tend to cover the axilla hence making them less anxious about the cosmetic factor. Women who wear sleeveless are more prone to report in view of the cosmetic factor. Accessory breasts are typically asymptomatic, but may also present with pain during the premenstrual phase. But they cause anxiety, cosmetic problems, pain or restriction of arm movement. It has been recommended that all accessory breasts be removed surgically. (4) Of the 95 accessory breasts that were operated on, in three years, six patients had carcinomas in them. Of these six, three patients have a rare type of cancer. Hence, we present three cases of accessory breasts, who had a rare type of carcinoma.
Case 1:
Thirty-four-year-old female presented with a left axillary swelling for the past 3 years. She had pain during the premenstrual period. Examination proved an accessory breast with a probable mass. FNAC only showed normal breast tissue. Ultrasound showed breast architecture with intra-lobular fluid. Hence, she was taken up for surgery, and excision was done. Histopathology proved an intra-cystic papillary carcinoma. (ER – Positive, PR – negative, HER 2neu – Negative). Hence, she was subjected to hormonal and chemotherapy post suture removal.

Figure 1: specimen of excised axillary accessory breast

Case 2:
Forty-one-year-old female presented with a left sided chest wall swelling since birth, which grew to the current size. Examination proved to be an accessory breast with a nipple and areola. She wanted it removed for cosmetic reasons. Post excision, histopathology reports suggested intra-cystic papillary carcinoma. (ER – negative, PR – negative, HER 2neu – Negative). Hence, she was subjected to chemotherapy post-suture removal.

Figure 2: accessory breast with NAC in the left chest wall

Case 3:
Fifty-two-year-old female came with a lump in the left axilla which was present only for the past 2 months. Examination revealed an axillary mass. Investigations proved breast cancer with axillary nodes. Hence, she was taken up for modified
radical mastectomy. Histopathology suggested intra-cystic papillary carcinoma of the accessory breast with 2 axillary lymph nodes. Her breast was normal according to pathology. (ER – positive, PR – positive, HER 2neu – Negative). Hence, she was later subjected to hormonal and chemotherapy.

Discussion

Papillary carcinoma occurring in the breast is a rare malignant tumour. It occurs in about 1-2% of all breast carcinomas. (5) It usually has a papillary structural design, similar to that of a finger like projection composed of central fibrovascular core covered by epithelium, without myoepithelial cell layer (difference between benign and malignant). (6) It may be invasive or non-invasive. Non-invasive type can be further subdivided into two subtypes: a diffuse form (the papillary variant of ductal carcinoma in situ) and a localized form (intra-cystic papillary carcinoma). The localized form, is usually present as a solitary tumour in an encysted or dilated duct. This is more common among postmenopausal women. it usually presents as benign mass. In some cases bloody nipple discharge may be there and it might also be asymptomatic. Axillary nodes are not common. (7,8) Mammography reveals round, ovular, or lobulated opacity with indistinct margins either due to invasion or inflammation. Differential diagnosis for this includes hematoma, invasive ductal carcinoma, and colloid or medullary carcinoma, benign cyst, or adenofibroma. (9,10)

Ultra sound may show an indistinct border or microlobulation, suggestive of malignancy, it may show septations with papillary projections. These may often be related to spontaneous haemorrhage within the tumour in the cyst. (9,11) Fine needle aspiration cytology may show a frankly bloody aspirate. Core needle biopsy has a low accuracy even while combined with ultra sound guidance in distinguishing in-situ and invasive carcinomas. This is because the site of biopsy is generally central while the invasion is usually found in the periphery of the tumour. (7,8,12)

Literature states that an excision biopsy is sufficient if it involves only the accessory breast, a sentinel lymph node biopsy can be done in order to review the need for a full-blown axillary dissection. However, diagnosis is usually histological, hence there might be a radical surgery done in view of cancer clearance. (13–17) The largest study was done from the database of the Californian cancer registry. The study was from 1988 to 2005. An analysis of over 900 cases was done. It demonstrates that distinguishing between in situ or
invasive intra-cystic papillary carcinoma is not of clinical significance because of the excellent prognosis. (18)

So far there have been no evidence-based guidelines for the treatment of intra-cystic papillary carcinoma and no randomised controlled trial comparing different types of surgeries. There have been many cases reports and case series showing excellent prognosis with conservative management. Axillary dissection has no association with prognosis. Hence, a sentinel lymph node biopsy can be done prior to an axillary dissection. (17, 19, 20)

**Conclusion**

Cancer of the accessory breast is rare. And carcinomas occurring in them are ever rarer. Intra-cystic papillary carcinoma is slow growing with an excellent prognosis. The 10-year survival rate is reported to be 100%. Segmental mastectomy is usually performed, and sentinel lymph node biopsy is suggested.

**References**