Ectopic Breast Carcinoma

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ABSTRACT
A 47-year-old premenopausal woman had a swelling in right axilla which had been diagnosed as “ectopic breast tissue” with an incisional biopsy. A subcutaneous nodule appeared two years ago. The ectopic breast containing the mass was excised with axillary dissection. It was an invasive ductal carcinoma and metastasis was detected in one lymph node. She received local radiotherapy after 6 cycles of chemotherapy and has now been taking hormonotherapy. Ectopic breast tissue has potential for malignant transformation. As its carcinoma has a worse prognosis and a higher incidence of metastasis because of delayed diagnosis, prophylactic excision may be recommended.

Key words: Ectopic breast cancer. Breast carcinoma. Axillary breast.

INTRODUCTION
Milk lines are bilateral ectodermal ridges extending from anterior axillary folds to inguinal folds. During embryogenesis, they regress, except for the thoracic region, where they form the breasts. When the regression fails, ectopic breast tissue (EBT) develops and axilla is the most common site of localization.1,2 Although EBT may contain a combination of breast parenchyma, areola and nipple, the latter two are very seldom and the diagnosis is sometimes difficult. EBT usually becomes clinically prominent with swelling and tenderness during puberty, pregnancy and lactation.2 Unless it causes symptomatic or cosmetic discomfort, management is usually conservative. However, EBT, when undiagnosed with cytology, may cause confusion in management of patients with breast carcinoma, and pathological changes observed in normal breasts, such as fibroadenoma, fibrocystic changes, papilloma and ductal hyperplasia, can also develop in EBT.1-4 Ectopic breast carcinoma (EBC) is rarely observed and, because unsuspected, detection is usually late causing poorer prognosis for the patient.

This case report describes a case of ectopic breast carcinoma treated with excision and axillary dissection.

CASE REPORT
A premenopausal woman, 47 years of age, applied to the ambulatory unit for breast cancer screening in February, 2008. Ten years ago she had an incisional biopsy from a 50 mm-sized swelling at her right axilla. Pathological examination revealed an EBT. In January 2006, she realized that the skin at the location of the incisional biopsy got harder but it was accepted as fibrosis by the physician she consulted.

On current examination, the patient had a transverse biopsy scar of 15 mm-length on an ectopic breast tissue of 80 x 60 mm size at right axilla. Just behind the scar there was a hard nodule of about 1 cm which was fixed to both the skin and underneath. On ultrasonography, the mass was heterogenous, hypoechogenic and solid with size of 9.4x7.2 mm. There were 3 lymph nodes with the hilum preserved in the right axilla and the biggest one was 16 mm in diameter. On mammography, axillary mass looked malignant because of the cluster of microcalcifications inside and the spicular elongations around (Figure 1). No additional lesion was observed on MRI. Ultrasonography-guided needle biopsy of the axillary mass showed malignancy. The axillary breast tissue containing the malignant mass was excised with level 1 and 2 lymph node dissection. The 90x70x30 mm-sized tissue excised with uppermost skin from axilla was breast tissue. On sectioning there was a 10 mm-sized, hard and dirty-white colored mass with irregular borders just under the skin (Figure 2). On microscopic examination, epidermis was regular. Tumour had originated at the junction of dermis and subcutaneous fatty tissue and extended by infiltration into subcutaneous fat (Figure 3). Architectural pattern was mostly solid but adenoid at some places. In the neighborhood of the tumour skin, appendages with apocrine and eccrine differentiation, and sinusoidal and ductal structures of normal breast were visible. Just at the vicinity of the tumour there were areas of in-situ ductal carcinoma of solid and cribriform patterns of intermediate grade (Figure 4). On immunohistochemical examination, tumour cells were stained strongly for

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**DISCUSSION**

EBT is observed with an incidence of 2-6%. In about two-thirds of the cases it is located on axilla whereas most of the remainders are on vulva or the chest wall. Axillary EBT is usually in a superficial position and it is necessary to observe mammary glandular tissue interspersed among adnexal glands of the skin in order to differentiate it from axillary tail of eutopic breast.

EBC, however, forms about 0.3% of all breast cancers. Carcinoma might develop due to stagnation in ductal lumens after hormonal stimuli. Most common type is ductal carcinoma, but medullary, mucinous, papillary and lobular carcinomas and cystosarcoma phylloides have also been reported.

Clinically, differential diagnosis of EBC includes lipoma, enlarged lymph node, sebaceous cyst or hyalroniditis suppurativa and thus it is usually not diagnosed preoperatively. Sonographic examination usually rules out a lipoma and a lymph node but sometimes incorrectly diagnoses the hypoechoic mass close to the skin as a sebaceous cyst or hyalroniditis.

In this case, the nodule was very close to skin, hard and fixed. It could easily be thought of as a sebaceous cyst or a reaction to a thread but radiological examination was diagnostic for malignancy. On retrospective evaluation of the mammograms taken one year ago, it was realized that the carcinoma of a smaller diameter was visible.

Pathological diagnosis of EBC may be difficult, as well. In presence of a tumour accompanying normal apocrine ducts that is located in the junction of dermis and subcutaneous fatty tissue, and that is infiltrative with solid and tubular architecture, differential diagnosis must include invasive ductal carcinoma originating from axillary tail of the breast, carcinoma of skin appendages and invasive ductal carcinoma originating from EBT. For the diagnosis of primary tumour of EBT, benign breast tissue adjacent to tumour must be seen in the specimen. Presence of in situ carcinoma around the tumour and receptor positivity may also help. Eccrine sweat gland carcinomas lack staining for GCDFP-15 compared to ductal carcinomas of the breast. Also, in contrary to metastatic breast carcinomas, primary sweat gland carcinomas are positive for epidermal growth factor receptor. Several other immunohistochemical markers, such as S100, carcinoembryonic antigen, and c-erbB-2 oncoprotein have been found to be less helpful in differential diagnosis.

Prognosis is reported to be the same as that of eutopic breast carcinoma of the same stage, and lymphatic involvement is the most important prognostic factor. In ectopic axillary breast carcinoma lymphatic spread is more common (incidence, 59-88%) than superior outer quadrant tumours of the breast. This may be due to either early metastatic potential, its location, or delayed diagnosis. Follow-up data of 90 EBC patients revealed a combined survival of 9.4% beyond 4-year post-treatment period. However, most of the patients in this study were diagnosed at TNM stages of III or IV. Prognosis will be better if the diagnosis is made early in the course of the disease. Thus, suspicion of malignancy in an EBT by the surgeon is an important first step to make the diagnosis which will prevent delays in treatment.

For EBC wide local excision and axillary node dissection is the optimal treatment. Sentinel lymph node biopsy can be performed if technically possible. Modified radical mastectomy has no additional survival advantage if the breasts have no additional lesions. Of the 28 cases that survived, 12 had recurrence in the series of Evans et al. and thus careful follow up of the patients is necessary. In this case, local excision of the EBT containing the carcinoma was performed with free...
surgical margins. As there was a palpable lymph node which looked malignant, we performed formal axillary dissection instead of sentinel node biopsy.

Adjuvant therapies for EBC must also be planned according to the current protocols prepared for eutopic breast carcinomas.\textsuperscript{2,6,10} Local control with radiotherapy may be necessary because of higher recurrence rates observed after removal of EBC. Tamoxifen is reported to be effective in prevention of cancer recurrence also in patients with EBC when the tumour is hormone-responsive.\textsuperscript{1} We also started hormonotherapy with tamoxifen as the tumour was strongly positive for receptors.

Prophylactic removal of EBT before the development of carcinoma has been discussed but is still controversial. Authors defending prophylactic excision claimed that prognosis of EBC was worse than that of normal breast carcinoma.\textsuperscript{10} However, it was reported that the worse prognosis of EBC was not due to the more aggressive behaviour of the tumour but usually because of late diagnosis. Also, EBC was said to form less than 1\% of all breast cancers although the incidence of EBT was more common.\textsuperscript{5}

However, several aspects of carcinoma of EBT may justify prophylactic resection. It is reported that EBT is more prone to malignant transformation than normal breasts and that EBC can metastasize like normal breast carcinoma which may be earlier, especially if localized in axillary region.\textsuperscript{10}

Most patients with EBT have complaints, both cosmetic and symptomatic. As the surgeon will not enter the axillary cavity when removing the superficially located EBT, prophylactic surgery does not result in complications like lymphedema or chronic arm pain. Thus our recommendation is to talk to the patient about the risk of malignant changes and excise the ectopic breast tissue. If the patient prefers a conservative approach without surgery, axillary region should be included in the radiological examinations on follow-ups and any change in axilla should alarm the patient for consulting a breast surgeon.

EBC is seen seldom but, when diagnosed, it is usually in an advanced stage which results in a worse prognosis. A high suspicious approach to masses in axilla, especially in the presence of EBT, will decrease the incidence of missed diagnosis. Although controversial in literature, prophylactic excision of EBT, at least in patients with symptomatic or cosmetic discomfort, might be recommended.

**REFERENCES**


