Carcinoma Originating from the Ectopic Mammary Gland of the Axilla

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Key Words

Ectopic breast · Axilla · Carcinoma · Kuwait

Abstract

Carcinoma originating from ectopic breast tissue is a rare condition comprising only 0.3% of all breast cancers. The case presented here is the first of its kind to be reported in Kuwait. A review of the literature is provided, and a rationale for the treatment of this condition.

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Introduction

Ectopic breast tissue may occur anywhere along the embryonic milk line. Carcinoma of the axillary breast is a rare finding, accounting for approximately 0.3% of all breast cancers. A search of the literature showed that, to date, a total of 95 cases of carcinoma of ectopic

breast tissue have been reported worldwide, 69 of which occurred in the axilla [1–6].

The correct preoperative diagnosis is rarely made for this condition. We report a case of carcinoma originating from ectopic breast tissue of the axilla, and review the available literature. Wide local excision and axillary lymph node dissection are necessary [7]. Adjuvant chemotherapy and irradiation, together with endocrine therapy, improve the outcome [8, 9].

Case Report

A 49-year-old woman presented with a slowly growing painful lesion in the left axilla, which had been slowly growing for the past 2 years. Both the size of the swelling and the pain increased during her regular menstrual cycles. She was not on contraceptive pills or hormonal treatment. The lesion was 2.5 × 1.5 cm in diameter, hard in consistency, brown in colour with central punctum and attached to the overlying skin. Bilateral mammography and breast ultrasound ruled out any associated lesion. Fine-needle aspiration cytol-

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Fig. 1. Malignant lobular carcinoma cells in 'indian file pattern' surrounding skin adenexal glands of axilla. HE. $\times 400$.

Fig. 2. Cells of invasive lobular carcinoma showing intracytoplasmic lumina. HE $\times 800$.

ogy was benign. Under general anaesthesia, the patient underwent wide local excision together with axillary dissection. Frozen section was negative for malignancy. However, paraffin sections revealed invasive lobular carcinoma arising from ectopic breast tissue in the axilla (fig. 1, 2). The tumour diffusely infiltrated the densely fibrotic dermis and the subcutaneous tissue. The tumor did not invade the epidermis. Oestrogen receptors were positive, as determined by immunohistochemical examination. Resection lines were free of tumour. One out of 14 lymph nodes showed a tiny tumour deposit in its perinodal fat. Chest X-ray, liver ultrasound examination and bone scan were normal.

The patient received 6 cycles of adjuvant chemotherapy: cyclophosphamid, methotrexate, 5-fluorouracil (CMF) for a total of 6 courses, together with tamoxifen 20 mg daily for 5 years.

Radiation therapy for the internal mammary and supraclavicular lymph nodes was given upon completion of the CMF course. Regular follow-up up to 36 months revealed no clinical or radiological evidence of local recurrence or distant metastasis.

Fig. 3. Diagram of the milk line demonstrating sites of supernumerary nipples and breast tissue.

Embryological Characteristics

Breasts are skin appendages arising from ectodermally derived mammary lines in the embryo. This line extends from the forelimb to the region of the hind limb in the 7-mm embryo. Development of the breasts proceeds identically in both sexes during foetal life (fig. 3). Most of the mammary line disappears soon after its formation, but a small portion in the thoracic region persists and thickens to form the mammary primordium. The ectoderm of the primordium penetrates to the underlying mesenchyme and gives rise to 15–25 epithelial strips that, by the end of prenatal life, canalise and form the lactiferous ducts and the alveoli of the mammary gland. The primitive lactiferous duct system begins peripherally and proceeds towards the eventual nipple/areola complex. They begin by opening into an epithelial pit, which at term or later gives rise to the nipple as a consequence of proliferation of surrounding mesoderm.

Usually, the mammary line persists only in the thoracic region in the human, but occasionally residual fragments give rise to accessory mammary tissue. Accessory breast tissue may take several forms. It has been classified in the following manner: (1) complete breast with nipple, areola and glandular tissue; (2) supernumerary breast without areola, but with nipple and glandular tissue; (3) supernumerary breast without nipple, but with areola and glandular tissue; (4) aberrant glandular tissue without nipple and areola; (5) pseudomamma with nipple and areola, but without glandular tissue – the breast is replaced by fat; (6) polythelia – presence of a nipple only; (7) Polythelia areolaris – presence of an areola only, or (8) polythelia pilosa – presence of a patch of hair only.

While aberrant breast tissue usually develops somewhere along the embryonic mammary line, in rare instances it has appeared in other unusual sites such as the shoulder, buttocks or face. Supernumerary nipples can be identified at birth, whereas ectopic breast tissue becomes noticeable only after hormonal stimulation, usually during puberty, pregnancy or lactation.

Discussion

The mammary line persists only in the thoracic region in humans, but occasionally residual fragments give rise to accessory mammary tissue. Polymastia has been found in approximately 2–6% of women [10, 11], whereas the majority of women with polymastia have only 1 additional breast, 2 or more are not uncommon, and up to 10 additional pairs of breast have been reported [11]. Supernumerary breast tissue and nipples are often dismissed as cosmetic curiosities. These structures have the potential for pathologic degeneration and may be associated with significant

congenital abnormalities of the urinary and cardiovascular systems. Urbani and Betti [12] found a significantly higher frequency of kidney and urinary tract malformations in cases that had accessory mammary tissue. The affected patients compared to the controls were 7.53% versus. 0.68%, respectively [13]. A supernumerary nipple is also regarded as a cutaneous paraneoplastic marker because of its significant association with urogenital malignancy [13–16]. The incidence of carcinoma in an ectopic breast is rare [1, 8]. It is difficult to predict the prognosis since some authors suggested that the outlook might be worse than carcinoma occurring in pectoral breasts because of the earlier involvement of the draining lymph nodes [9]. Other reports showed long-term survival in node-negative patients [8, 17, 18]. Evans and Guyton [1] quote the work by Razemon and Rizard, who in 1927 collected a series of 17 cases of which 13 were treated by breast preservation and axillary

dissection and 4 by radical mastectomy. Ten of them recurred within 2 years of surgery, and 3 survived for less than 2 years. They concluded that additional, radical or modified radical mastectomy offered no advantage in outcome over axillary mastectomy and regional node dissection [9, 17, 18].

Conclusion

Pathologic changes are liable to develop in an axillary breast. This creates a diagnostic challenge. Awareness and high index of suspicion of extramammary swellings along the milk line that correlates with menstruation or pregnancy is strongly emphasized. Early wide excision of such lesions may improve survival.

Treatment policy should be similar to that advocated for breast cancer in general. Long-term follow-up is essential.

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