

The management of ectopic breast cancer - Case Report

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Summary

Ectopic breast tissue occurs when the mammary ridge fails to resolve during embryonic development. Ectopic breasts, either supernumerary or aberrant, have an incidence of 6%. Ectopic breast tissue is however hardly mentioned in the literature. Also benign and malignant alterations in these "breasts" are under-reported. The lifetime risk of women being diagnosed with breast cancer is 13%. It should be recommended that these "breasts" be included in the screening. Due to the unawareness this is generally not happening. The present article describes a case of ectopic breast cancer and discusses therapeutic management. A subcutaneous mass along the 'milk line' should be examined carefully and any suspicious lesions should be approached according to the guidelines for the management of symptomatic breast disease. Ipsilateral prophylactic mastectomy has no role in the management of a single ectopic breast cancer tissue.

Key words: Ectopic; Accessory; Aberrant; Breast; Cancer; Screening.

Introduction

Folk wisdom associated extra nipples and breasts with exceptional virility and fertility [1-3]. The Phoenician god of fertility Astarte had a plethora of breasts [3]. Also Darwin believed that this was an atavistic trait that supported the theory of evolution [1, 2, 4, 5]. There is however no scientific evidence for these beliefs.

Ectopic breast tissue is divided in the literature between aberrant and supernumerary breasts. The incidence of the first is unknown and for the latter it is estimated to range between 0.6% (Caucasians) and 6.0% (Japanese), depending on the ethnic population [6-8]. About 70% of ectopic breast tissue is localized in the axilla [6]. The majority are asymptomatic and women will only become aware of this tissue during hormonal changes like puberty, menstruation, pregnancy or lactation. The size of ectopic breasts can vary from small to the size of a normal breast [9]. However like the pectoral breast they are susceptible to all the changes of the reproductive cycle. In a series of 233 patients with an axillary accessory breast 66 underwent a surgical excision [10]. In 59% for cosmetic concerns, in 30% for mild pain, in 9% for cancer and in 2% for an abscess. Practically all breast diseases have been reported in ectopic breast tissue [11-14]. Breast cancer is the number one cancer in the developed world and affects about one in eight women. Only 0.3% of the reported breast cancers are of ectopic origin. In the literature there are less than 100 reports of axillary breast cancer. This is undoubtedly an underestimation. It is important to be aware of ectopic breast tissue and the possibility of malignant transformation. The present report will give an overview of the diagnosis and management of ectopic breast cancer.

Case Report

A 51-year-old woman was consulted for a painful nodule in the left axilla. Clinical examination showed an enlargement of tissue in the axilla with an overlying red subcutaneous lesion in the middle. The latter was suggestive of an infected sebaceous cyst with inflammation of the surrounding tissue. A screening mammography and ultrasound was within normal range. However the mammography demonstrated a moderately sized area of density in the left axilla that resembled glandular parenchyma, separate from the bulk of parenchyma. In the middle of this glandular tissue an ill-defined nodule 1 cm in diameter was seen. The clinical picture together with the mammography was suggestive of the presence of aberrant breast tissue in the axilla. Magnetic resonance imaging (MRI) was performed as a function of a research protocol. The MRI scan could also demonstrate this nodule and showed that the lesion had benign features. It was assumed that this lesion caused the pain and therefore, despite its benign characteristics, was excised. Histological examination demonstrated a moderately differentiated ductal adenocarcinoma 0.6 cm in diameter. The tumour was completely removed; it was positive for oestrogen and progesterone receptors. With this information, a mastectomy of the aberrant breast tissue and axillary lymph node dissection was performed (Figure 1). There was no residual carcinoma in the aberrant breast and all 14 lymph nodes were free of tumour. A search performed for occult metastases (chest X-ray, bone scan and liver ultrasound) was negative. After multidisciplinary deliberation it was decided to advise the patient to undergo adjuvant hormonal therapy (tamoxifen 20 mg daily) for five years. At present 51 months after the diagnosis the patient is well with no signs of recurrence.

Discussion

Ectopic breasts are anomalies of embryonic development. During the fourth to the fifth week of gestation a mammary ridge or milk line appears which extends from the axilla, over the nipple to the groin. A part of these cells will proliferate and lead to the mammary bud, and

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Figure 1. — Clinical presentation of axillary ectopic breast tissue with outlining of the mastectomy.

the rest of the ridge normally disappears. Failure of resolution of this mammary ridge accounts for the occurrence of ectopic breast tissue. Ectopic breast tissue has also been reported to occur in locations outside the milk line, including the face, posterior neck, chest, parasternal, subscapular, midback, buttock, vulva, flank, hip, posterior and lateral aspects of the thigh, shoulder, and upper extremities [15, 16].

In 1915 already Kajava proposed a classification for ectopic breast tissue and nipples [17]. Ectopic breast tissue can be classified as complete breast (polymastia), glandular tissue with papilla but no areola (supernumerary breast), glandular tissue with areola but no papilla (supernumerary breast), glandular tissue only (ectopic breast tissue), papilla and areola only with gland replaced by fat (pseudomamma), papilla only (polythelia), areola only (polythelia areolis) and patch of hair only (polythelia pilosa). In the literature ectopic breast tissue is generally divided in supernumerary and aberrant breasts. Where supernumerary breasts contain a nipple-areola complex, with or without a duct system that communicates with the overlying skin, aberrant breast tissue is considered an island of breast tissue in absence of a nipple or areola, primarily in the neighbourhood of the breast. The aberrant breast is mainly localised in the axilla, and less frequently the aberrant breast can be found in the infraclavicular, juxtosternal or epigastric region [16]. Histologically there is no organised secretor system and they do not bear any relationship to the overlying skin. There is also no histological connection with the pectoral breasts.

There appears to be a hereditary link. Polythelia is transmitted in an autosomal dominant trait with an incidence of 0.22-2.5% and it may be associated with renal anomalies like urogenital defects and cancer [2, 5, 16, 18-24]. Polythelia in males has been associated with testicular cancer [22]. Polymastia is associated with renal anomalies in 40%. Several combinations with chromosomal abnormalities (anomaly of chromosome 3 and 8, Turner syndrome, Fleischer's syndrome), vertebral anomalies, multiple endocrine neoplasias, ventricular defects and

pyloric stenosis have been reported in the literature [1, 7, 11, 19-21, 23, 24]. The coexistence of multiple endocrine neoplasia and ectopic breast tissue suggests a common embryonic ectodermal tissue origin [11, 25].

Accessory breast tissue should be approached in the same way as "normal" breast tissue - whether or not there is an indication to screen ectopic breasts is unknown. However given the high incidence of breast cancer we recommend it. In case of a suspicious lesion triple assessment (clinical examination, radiological evaluation and minimal invasive biopsy) should be done as in "normal" breast tissue.

An axillary mass evokes multiple diagnoses. These differential diagnoses can be divided in four categories: (1) malignancy (primary or secondary) (breast cancer, neuroendocrine tumour, epidermoid carcinoma, rhabdomyosarcoma, lymphoma, melanoma, metastasized cancer to axillary lymph nodes); (2) infectious (suppurative hidradenitis, cat scratch disease, lymphadenopathy, lymphadenitis tuberculosis); (3) vascular (lymphatic dilatation, pseudoaneurysm, aneurysm); (4) miscellaneous: axillary tail of Spence, torn muscle belly, sebaceous cyst, excess of axillary fat [26, 27].

Accessory breast tissue is normally not excised, except for cosmetic reasons. The management of ectopic breast cancer should follow the current recommendations for breast cancer. Although patients with ectopic breast cancer have an increased risk of developing a second primary tumor, this does not warrant an ipsilateral prophylactic mastectomy. The aberrant breast tissue should be completely removed via mastectomy or wide local excision of the accessory breast, and a lymphadenectomy should be performed based on the lymphatic drainage as defined by Sappey in 1885 [28]. TNM staging should be similar to breast cancer [29]. For the lymphadenectomy it is important to recognise that Sappey's line extends circumferentially from the inferior costal margin anteriorly to the inferior border of the 12th rib posteriorly and is 2 cm wide. Lesions above this line would preferentially drain to the axilla and those below this line preferentially to the inguinal region. The sentinel node mapping technique would probably be very helpful in identifying the preferable drainage location of the tumour.

Recently in one of the first descriptions on the use of sentinel node biopsy in the treatment of accessory breast cancer a positive intramammary lymph node location was identified [30]. This unusual lymph node group has not been previously described and could be part of the pathway to the internal mammary chain.

From the reported cases in the literature it seems likely that ectopic breast cancer has the same prognosis as non ectopic breast cancer when corrected for TNM [31]. The prognosis can only be improved by early detection. Therefore every nodule in a region of ectopic breast tissue should be approached with the same consciousness as in normal breasts. Unfortunately ectopic breast tissue without the presence of a nipple is frequently misdiagnosed as axillary fat, lymphadenitis, suppurative hidradenitis, lipomas or other soft tissue tumours [26,

27]. Accessory nipples on the other hand are often mistaken as nevi.

Women and doctors should be aware of the possibility of ectopic breast tissue in order to reduce anxiety. Ectopic breasts are as vulnerable to changes as "normal" breasts. Furthermore ectopic breasts should be included in radiological screening. The approach to a suspicious ectopic breast lesion(s) should be like the approach to non-ectopic breasts: triple assessment. The management of ectopic breast cancer should be similar to non-ectopic breast cancer.

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