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# **Breast Imaging**

# Inflammatory breast cancer in accessory abdominal breast tissue

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## ABSTRACT

Accessory breast tissue results from failure of the embryologic mammary ridge, also known as the milk line, to involute. As a result, ectopic breast tissue can develop anywhere along this ridge, which extends from the axilla—the most common location—to the groin. Primary breast cancer in accessory breast tissue is uncommon but has been reported in multiple prior studies. We present a rare case of inflammatory breast cancer presenting in upper abdominal accessory breast tissue in women with a personal history of ipsilateral breast cancer, and highlight the challenges of both diagnosis and treatment of breast cancer in accessory breast tissue.

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## Introduction

Accessory breast tissue results from incomplete involution of the embryologic mammary ridge, also known as the milk line. As a result, ectopic breast tissue can develop anywhere along this ridge, which extends from the axilla—the most common location—to the groin [1,2]. Primary breast cancer in accessory breast tissue is uncommon but has been reported in prior studies [3–5]. We present a rare case of inflammatory breast cancer presenting in upper abdominal accessory breast tissue in a woman with a personal history of ipsilateral breast cancer, and highlight the challenges of both diagnosis and treatment of breast cancer in accessory breast tissue.

## **Case report**

A 58-year old woman with a history of stage I left breast cancer (T1aN0M0) treated 9 years prior with breast conservation therapy, radiation, and hormone therapy presented with a 2-week history of localized erythema measuring 4 cm and tenderness involving the patient's known supernumerary nipple located in the left upper abdomen. The patient was treated with antibiotics over a 2-week period without improvement of symptoms. Subsequent targeted ultrasound demonstrated a microlobulated mass measuring 20 mm located within accessory breast tissue (Fig. 1A). Ultrasound-guided biopsy of this mass was performed with postbiopsy mammogram demonstrating a biopsy marker clip at site in the mass within

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Fig. 1 – (A) Targeted left abdominal accessory breast ultrasound demonstrates a microlobulated 20-mm mass with overlying skin thickening. (B) Post-biopsy left mammogram including the abdominal accessory breast, left lateromedial view (LLM), demonstrates a mass with a biopsy marker clip at site (white arrow). (C) Hemotoxylin and eosin staining of the ultrasound-guided biopsy sample showed an invasive ductal carcinoma with suspicious lymphovascular invasion (black arrow).

accessory breast tissue (Fig. 1B). Hematoxylin and eosin staining of the biopsy sample showed invasive ductal carcinoma (IDC) with suspicious lymphovascular invasion (Fig. 1C). The tumor was estrogen receptor positive (ER+), progesterone receptor negative (PR+), and Her2/neu receptor negative. The metastatic workup with computed tomography chest, abdomen, and pelvis and positron emission tomography-computed tomography was negative. Because clinical findings were consistent with inflammatory breast cancer, the patient began neoadjuvant chemotherapy resulting in resolution of skin changes. After the completion of neoadjuvant chemotherapy, the patient underwent a surgical resection with final pathology revealing grade 3 IDC, estrogen receptor positive, progesterone receptor negative, Her2/neu receptor negative, 60% cellularity, associated intermediate- to high-grade DCIS, and suspicious lymphovascular invasion. Surgical margins were negative (>5 mm) and 1 sentinel lymph node was negative for malignancy. Following surgical therapy, the patient underwent targeted radiation therapy with capecitabine (Xeloda) and was started on a 10-year course of anastrozole (Arimidex).

## Discussion

Accessory breast tissue develops from incomplete embryologic regression of the mammary ridge, typically extending from the axilla to the groin. Although the axilla represents the most common location, ectopic mammary tissue can also develop in the face, posterior neck, shoulder, chest, middle back, buttock, perineum, vulva, hip, or groin [6,7]. Accessory breast tissue consists of any combination of breast parenchyma, areola, and nipple, and is susceptible to the same stimuli as native breast tissue, often becoming evident during puberty, pregnancy, or lactation [8]. Overall, the incidence of accessory breast tissue in the general population ranges from 2.0% to 6.0%, occurring in both men and women [9–11]. Although most cases are sporadic, inheritance may be autosomal dominant with incomplete penetrance [12].

Primary breast carcinoma in accessory breast tissue is a rare phenomenon, representing 0.2%-0.6% of all breast cancers [13–17]. Although there is no established age predilection, prior reports have demonstrated a 37% incidence in women under

the age of 45 years [18]. Clinical diagnosis is often delayed, however, due to physician unfamiliarity with the condition. The ectopic location of accessory breast tissue, which may lack an overlying areola or nipple, provides an additional barrier to early diagnosis. The most common presentation is palpable abnormality along the milk line. It may additionally present as skin thickening, tenderness, and edema at the site of accessory breast tissue [19]. The axilla is the most common site of primary breast carcinoma in accessory breast tissue, representing up to 70% of all ectopic breast carcinomas followed by parasternal, subclavicular, submammary, and vulvar locations [2,8]. Increased incidence in the axilla is largely due to increased occurrence of accessory breast tissue in the axillary region compared with other sites.

Imaging findings are identical to primary breast cancer in the anatomic breast, if visualized by mammogram or breast ultrasound. Differential diagnosis of suspicious imaging findings in ectopic mammary tissue includes fibroadenoma, follicular cyst, lipoma, lymphadenitis, lymphoma, and metastatic carcinoma [20,21]. Pathologic diagnosis via core biopsy or gross excision is recommended to further evaluate suspicious imaging or clinical findings [22]. Pathologic diagnosis of malignancy within normal breast parenchymal tissue, separate from the proper mammary gland, is required to make the diagnosis of primary breast cancer in accessory breast tissue. Similar to breast cancer in the anatomic breast, the most common type of accessory breast tissue carcinoma is IDC [23].

Treatment of primary breast cancer in accessory breast tissue is also similar to primary cancer in the anatomic breast and may include any combination of surgery, chemotherapy, radiation, and endocrine therapy. Wide resection of the tumor and surrounding tissue including the skin and regional lymph nodes is the current standard of care [24]. Adjuvant chemotherapy and targeted radiation therapy to accessory mammary tissue is also recommended for local control in patients with locally advanced disease. Patients with a history of breast cancer in the anatomic breast, treated with breast-conserving therapy and radiation, and presenting with a breast cancer in ectopic breast tissue may be eligible for radiation, if the area covered is outside the prior radiation field.

Overall, treatment regimens are principally based off of primary therapy in the anatomic breast. Thus, our patient, who was suspected of having inflammatory breast cancer, was treated with neoadjuvant chemotherapy before surgical resection. Ipsilateral mastectomy of a nondiseased native breast or ipsilateral axillary lymph node dissection in addition to local excision remains controversial, as no survival benefit has been demonstrated in these practices compared with local excision alone [25]. Radiation therapy to a nondiseased ipsilateral anatomic breast is not recommended [26]. If additional disease is found in the anatomic breast or lymphatic system, however, standard primary breast cancer treatment would be required at that location [27].

Prognosis of accessory breast carcinoma is difficult to ascertain due to small sample sizes and limited follow-up. Poor outcomes compared to native breast cancer have been reported, however, which may relate to delayed diagnosis often until clinical symptoms are present [23]. A higher incidence of lymph node involvement compared with primary breast carcinoma in the native breast may also contribute to differential outcomes [18]. The location of primary breast carcinoma in axillary breast tissue closer to axillary lymph nodes has been linked theoretically to earlier nodal metastatic disease [26,28].

Clinical findings, including palpable abnormalities and skin changes along the mammary ridge, should prompt consideration of primary breast carcinoma in accessory breast tissue, especially if ectopic mammary tissue is known to be present. Early recognition by clinicians is necessary to prevent delayed diagnosis and unnecessarily extensive therapy.

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