Cystic Presentation of High-Grade Ductal Carcinoma In Situ in an Inframammary Accessory Nipple

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PRACTICE **POINTS**

- Accessory breasts (also referred to as ectopic breast tissue) develop when breast tissue is retained along the mammary ridge outside of the usual pectoral regions.
- Because accessory breasts may contain the same structures as anatomically correct breasts, they can be subject to the same benign or malignant changes.
- Clinical and pathologic correlation is prudent when interpreting ectopic mammary tissue, as various benign or malignant neoplasms may arise in this setting, especially if there are underlying genetic aberrancies or genodermatoses.

To the Editor:

The term *ectopic breast tissue* serves as an umbrella term that encompasses breast tissue positioned in anatomically incorrect locations, including the subtypes of supernumerary and aberrant breasts.[1] However, the more frequently used term is *accessory breast tissue* (ABT).[1] Supernumerary breasts have diverse variations of a nipple, areola, and/or ductal tissue and can span in size from a small mole to a fully functioning breast. This breast type maintains structured ductal systems connected to the overlying skin and experiences regular changes during the reproductive cycle. In contrast, an aberrant breast is isolated breast tissue that does not contain organized ductal systems.[1] Accessory breast tissue is prevalent in up to 6.0% of the world population, with Japanese individuals being the most affected and White individuals being the least affected.[1]

Accessory breasts typically are located along the milk line—the embryologic precursor to mammary glands and nipples, which extend from the axillae to the groin and regress from the caudal end spanning to the groin.[2] For this reason, incomplete regression of the mammary ridge results in ABT, most commonly in the axillary region.[3] Accessory breast tissue usually is benign and is considered an anatomical variant; however, because the histomorphology is similar to mammary gland tissue, accessory breasts have the same proliferative potential as anatomically correct breasts and therefore can form fibroadenomas, cysts, abscesses, mastitis, or breast cancer.[4] Accessory breast carcinomas comprise 0.3% to 0.6% of all breast malignancies.[5] Certain genodermatoses (ie, Cowden syndrome) also may predispose patients to benign or malignant pathology in ABT.[6] We present a rare case of accessory breast cancer in the inframammary region masquerading as a cyst. These findings were further supported by ultrasonography and mammography.

A 45-year-old White woman presented to our clinic for removal of a dermal mass underlying a supernumerary nipple at the left inframammary fold. Her medical history was noncontributory and was only remarkable for uterine fibroids. She developed pain and swelling in the

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left breast 1 year prior, which prompted her to seek medical attention from her primary care physician. Diagnostic mammography was negative for any concerning malignant nodules, and subsequent *BRCA* genetic testing also was negative. Six months after the diagnostic mammography, she continued to experience pain and swelling in the left breast and was then referred for diagnostic ultrasonography; 2 masses in the left breast suspected as infected cysts with rupture were identified (Figure 1). She was then referred to our dermatology clinic for evaluation and surgical extirpation of the suspected cyst underlying the accessory breast. The area subsequently was excised under local anesthesia, and a second similar but smaller mass also was identified adjacent to the initial growth. Dermatopathologic examination revealed an estrogen receptor– (Figure 2A) and progesterone receptor–positive (Figure 2B), *ERBB2 (HER2/neu)*–negative, nuclear grade III ductal carcinoma in situ (Figure 3).

Various ABT classification methods have been proposed with Brightmore[7] categorizing polymastia into 8 subtypes: (1) complete breast; (2) glandular tissue and nipple; (3) glandular tissue and areola; (4) glandular tissue only; (5) nipple, areola, and fat; (6) nipple only; (7) areola only; and (8) patch of hair only. De Cholnokey[8] focused on axillary polymastia, dividing it into 4 classes: (1) axillary tumor in milk line without nipple or areola; (2) axillary tumor with areola with or without pigmentation; (3) nipple or areola without underlying breast tissue; and (4) complete breast with nipple, areola, and glandular tissue. Fenench's[9] method is preferred and simply describes ABT as 2 subtypes: supernumerary and aberrant.[1,2,10] One study observed 6% of ABT cancers were the supernumerary type and 94% were the aberrant type.[1] Ductal lumen stagnation increases the risk for accessory breast carcinoma development.[10] Men have a higher prevalence of cancer in ABT compared to anatomically correct breast tissue.[11]

There currently is no standardized guideline for ABT cancer treatment. The initial clinical impression of cancer of ABT may be misdiagnosed as lymphadenopathy, abscesses, or lipomas.[12] The risk for misdiagnosis is higher for cancer of ABT compared to normal breast tissue and is associated with a poorer prognosis.[1] Despite multiple screening modalities, our patient's initial breast cancer screenings proved unreliable. A mammogram failed to detect malignancy, likely secondary to the area of concern being out of the standard imaging field. Ultrasonography also was unreliable and led to misdiagnosis as an infected sebaceous cyst with rupture in our patient. Upon review of the ultrasound, concerns were raised by dermatology

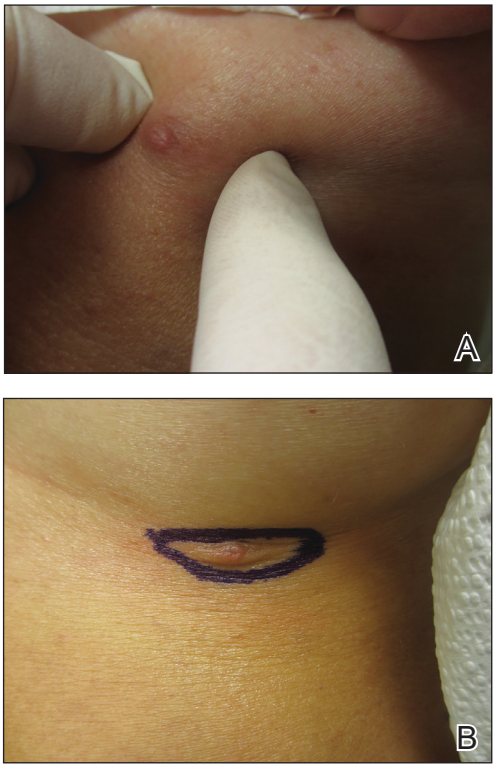

FIGURE 1. Gross clinical presentation of ductal carcinoma in situ. A, A 0.7-cm lesion at the 6-o'clock position of the left breast (7 cm from the nipple). B, A complex 1.7-cm mass accompanied by a 0.6-cm intradermal mass at the 6-o'clock position, 9 cm from the nipple along the left inframammary fold.

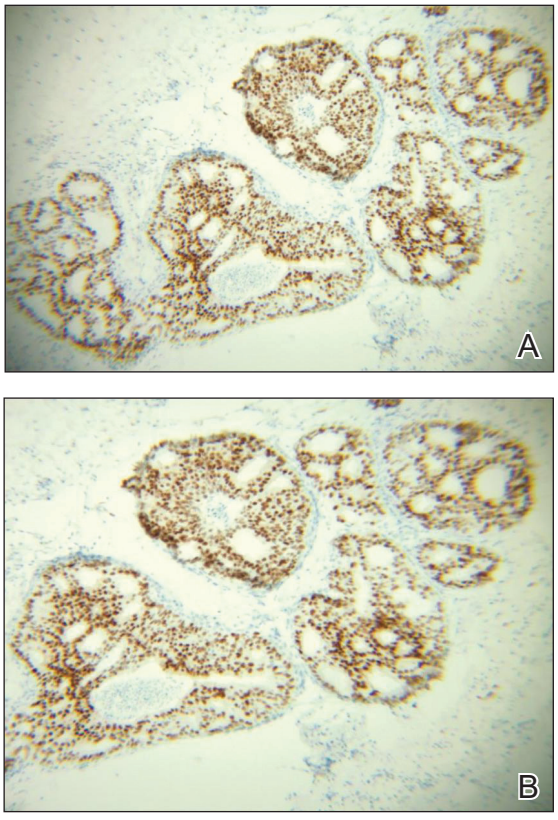

FIGURE 2. A and B, Immunostaining showed positive expressions of estrogen and progesterone receptors, respectively (original magnifications ×100).

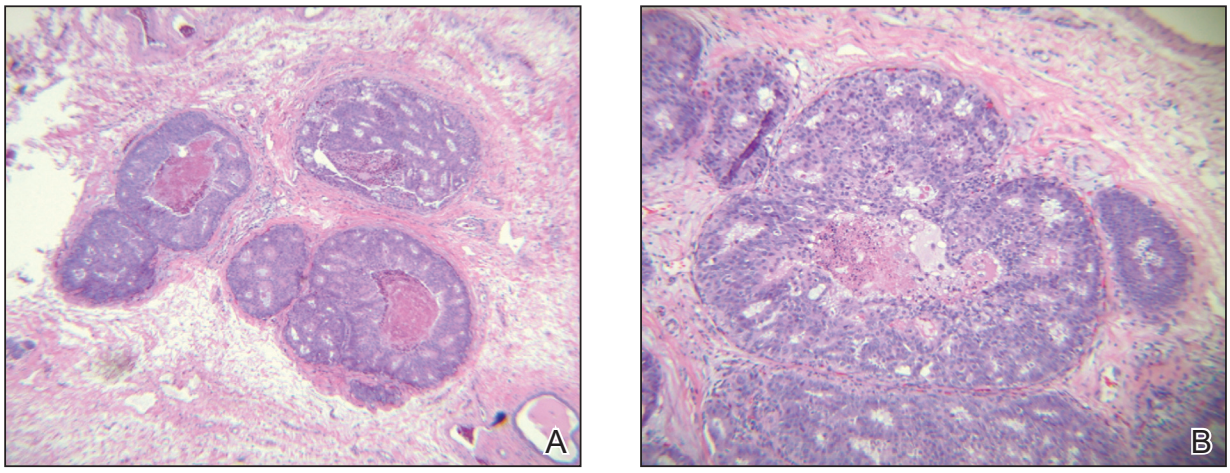

FIGURE 3. A, Histopathology revealed central expansile necrosis containing cellular debris, which generally is associated with high-grade ductal carcinoma in situ. There was fenestrated proliferation with multiple round, rigid, extracellular lumens with a punched-out appearance, distributed roughly equidistant and polarized, exhibiting a cribriform architectural growth pattern (H&E, original magnification ×40). B, Prominent pleomorphism and large nuclei (>2.5 times the size of a normal ductal epithelial cell) were seen. Cytologically, there was vesicular chromatin with irregular distribution, prominent nucleoli, and frequent mitoses. Cellular necrosis was present (H&E, original magnification ×100).

that the mass was more likely an epidermal inclusion cyst with rupture given the more superficial and sac-free nature of sebaceous cysts, which commonly are associated with steatocystoma multiplex.[13] Definitive diagnosis of ductal carcinoma in situ was made with dermatopathologic examination.

Prophylactic surgical excision of ABT has been recommended, suggesting that excisional biopsy and histopathologic examination is the more appropriate method to rule out malignancy. Surgical treatment of ABT may omit any risk for malignant transformation and may provide psychological relief to patients for aesthetic reasons.[10,12,14] The risk and benefits of prophylactic excision of ABT has been compared to prophylactic mastectomy of anatomically correct breasts,[15] with some clinicians considering this definitive procedure unnecessary except in high-risk patients with a strong genetic predisposition.[16,17]

Accessory breast tissue should be viewed as an anatomical variant with the option of surgical removal for symptomatic concerns, such as firm nodules, discharge, and pain. Although ABT is rare and cancer in ABT is even more uncommon (<1% of all breast cancers),[5,11] clinicians should be suspicious of benign diagnostic reports when the clinical situation does not fit the proposed narrative.

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