Pure acinic cell carcinoma of the breast
A case report and literature review
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Abstract
Rationale: Acinic cell carcinoma (AcCC) of the breast is a rare histological type of malignant epithelial neoplasm exhibiting acinic cell differentiation.

Patient concerns: A 52-year-old woman presented to the hospital with a palpable mass over the upper outer quadrant of the right breast.

Diagnoses: Physical examination revealed an irregular mass in the lateral upper quadrant of the left breast, approximately 1.5 cm in diameter. Histologically, the tumor cells were round to oval, had displaced nuclei with striking single nucleoli and basophilic cytoplasm, and contained large coarse cytoplasmic granules. They showed an infiltrating growth pattern with a combination of cystic and cribriform feature. We diagnosed the tumor as AcCC of the breast.

Interventions: The patient was given a simple mastectomy and sentinel lymph node biopsy. After the surgery, AcCC of the breast was confirmed histologically.

Outcomes: The patient was symptom free 3 months after surgery.

Lessons: AcCC of the breast is a very rare tumor, and its prognosis appears to be good. Thus, treatment followed the guidelines for invasive breast carcinoma and no further therapy was suggested by oncologists based on the tumor biology.

Abbreviations: a-1-ACT = a-1-antichymotrypsin, AcCC = acinic cell carcinoma, GCDFP-15 = gross cystic disease fluid protein 15, PAS-D = periodic acid–Schiff-diastase.

Keywords: acinic cell carcinoma, breast tumor, prognosis

1. Introduction

Acinic cell carcinoma (AcCC) of the breast is a rare histological type of malignant epithelial neoplasm exhibiting acinic cell differentiation. This type of neoplasm is commonly found in the parotid gland and has a diffuse infiltrative growth pattern, with small acinar or glandular structures composed of a monotonous proliferation of cells with a granular or clear cytoplasm, thus resembling the acinar cells of the salivary glands\textsuperscript{[1,2]} Since Roncaroli et al\textsuperscript{[3]} described the first case of AcCC of the breast in 1996, several cases have been reported in the literature.

In this article, we report a case of pure cystic AcCC of the breast in a 52-year-old woman, and comprehensively review similar cases of breast AcCC reported in the English-language literature to date.

2. Case presentation

A 52-year-old woman reported a 4-year history of a palpable mass over the upper outer quadrant of the right breast. Physical examination revealed an irregular mass in the lateral upper quadrant of the left breast, approximately 1.5 cm in diameter. No dimpling or palpable axillary or supraclavicular lymph node was detected. Results of laboratory tests were all within the reference ranges.

Grossly, the specimen was a 1.5 × 1.0 × 1.0-cm white-gray tumor with a well-defined border. The cut surface showed predominantly solid and small cystic areas, with visible hemorrhage. Histologically, the tumor cells were round to oval, had displaced nuclei with striking single nucleoli and basophilic cytoplasm, and contained large coarse cytoplasmic granules. Many had bright red granules resembling acinar cell zymogen granules. High-power viewing of glandular structures showed colloid-like secretions in the central lumina and cytoplasmic eosinophilic granules. They showed an infiltrating growth pattern with a combination of cystic and cribriform features (Fig. 1). The nuclear grade of the tumor cells was determined to be grade 1. The mitotic count was low among the malignant cells. No vascular or perineural invasion was observed. The sentinel lymph node was free of metastasis. The patient was symptom free 3 months after surgery.

Immunohistochemically, the tumor cells were positive for a-1-antichymotrypsin (a-1-ACT), cytokeratin 7, epithelial membrane
antigen, S-100 protein, gross cystic disease fluid protein 15 (GCDFP-15), and Periodic acid–Schiff-diastase (PAS-D) stain positivity (Fig. 2). The cells were partially positive for estrogen receptor and progesterone receptor, and were negative for human epidermal growth factor receptor 2, lysozyme, P63, smooth muscle actin. From the above results, we diagnosed the tumor as AcCC of the breast.

2.1. Ethics approval and consent to participate
This study was approved by the ethics committee of the Women’s Hospital, School of Medicine Zhejiang University.

2.2. Consent for publication
Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

2.3. Availability of data and material
The datasets during and/or analyzed during the current study available from the corresponding author on reasonable request.

3. Discussion
All breast AcCCs described in the literature since 1996 have affected women, most frequently in the sixth decade of life, and have presented as palpable nodules measuring 2 to 5 cm, with the right side being more commonly affected. The prognosis of AcCC of the breast appears to be good, even with reports of recurrence and metastasis.[4–6] Diagnoses have been based on morphological and immunohistochemical features.

The histological pattern of these cases is predominantly solid with a microglandular structure, with associated cystic or/and papillary components in 4 cases.[7–9] Interestingly, the present case showed an infiltrating growth pattern with cystic features. The cystic structures consisted of round cells with finely granular, weakly basophilic, or clear cytoplasm, resembling that of acinar cells of salivary glands.
AcCC is characterized by serous acinar differentiation with zymogen-type cytoplasmic granules. The immunohistochemical profile of breast AcCC shares many features with AcCC of the salivary glands, with frequent expression of GCDFP-15, S-100, and α-1-ACT, as well as PAS-D positivity. However, our case was negative for lysozyme, whereas most previously reported cases were positive for lysozyme\(^{10}\); 1 reported case was negative.\(^{11}\)

Forty-eight cases of breast AcCC have been reported in the English-language literature, including the new case reported in this article. Detailed morphological descriptions were provided for 32 of these cases.\(^{12}\) Many studies, based on very small case series, have suggested that breast AcCC is likely to have a good prognosis.\(^{3,13}\) The treatment approach seems to be similar to that for invasive breast cancer, but the impact on prognosis is not clear.

Six patients developed metastasis to sites such as the bone, liver, and lung\(^{[13–16]}\); 2 of these patients died of the disease.\(^{[14,15]}\) However, both patients who died had associated invasive ductal carcinoma not otherwise specified, and only single focal areas of AcCC positivity were noted. In 1 case, the invasive carcinoma had various morphological features, including solid areas of poorly differentiated carcinoma cells, areas of glandular differentiation typical of ductal carcinoma, and focal matrix-producing and adenoid cystic carcinoma-like areas.\(^{[14]}\) In the other case, a grade 3 infiltrating ductal carcinoma was revealed. The cells showed prominent nuclear pleomorphism with numerous mitotic figures.\(^{[13]}\)

4. Conclusion

Although metastasis and recurrence occur, their rates are so low that the prognosis of pure AcCC of the breast can be considered to be good. We followed guidelines for invasive breast carcinoma, and no further therapy was suggested by oncologists according to tumor biology.

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