Carcinoma adenoide cístico de mama: relato de caso de uma rara neoplasia

Adenoid cystic carcinoma of the breast: a case report of a rare neoplasm

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RESUMO

O carcinoma adenoide cístico representa aproximadamente 0,1% dos carcinomas da mama e corresponde a uma neoplasia de baixo grau de malignidade constituída por dois tipos celulares distintos (basaloides/mioepiteliais e ductais/epiteliais). Relacionado com o padrão molecular dos tumores de fenótipo basal, este carcinoma geralmente revela imunoexpressão negativa para os receptores hormonais e HER-2. No presente relato, os autores descrevem as características clinicas e histopatológicas de um carcinoma adenoide cístico originado no quadrante superior lateral da mama direita em uma paciente de 50 anos.

UNITERMOS: *Mama, Carcinoma Adenoide Cístico, Patologia, Imuno-histoquímica, Carcinoma Ductal Infiltrante.*

ABSTRACT

Adenoid cystic carcinoma represents approximately 0.1% of breast carcinomas and corresponds to a neoplasm of low grade malignancy composed of two different cell types (basaloid/myoepithelial and ductal/epithelial). Related to the molecular pattern of basal phenotype tumors, this cancer usually reveals negative immunostaining for hormone receptors and HER-2. In this report the authors describe the clinical and histopathologic features of an adenoid cystic carcinoma originated in the upper side of the right breast in a 50-year-old patient.

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KEYWORDS: Breast, Adenoid Cystic Carcinoma, Pathology, Immunohistochemistry, Invasive Ductal Darcinoma.

INTRODUCTION

Invasive breast carcinoma is the most common malignant neoplasia in women. It accounts for 22% of all female cancers. The vast majority of these tumors are adenocarcinomas and are believed to be derived from the mammary parenchymal epithelium, particularly cells of the terminal duct lobular unit. Breast carcinomas exhibit a wide range of morphological phenotypes and specific histopathological types have particular prognostic or clinical characteristics (1, 2, 3).

Adenoid cystic carcinoma represent about 0.1% of breast carcinomas. It is a carcinoma of low aggressive potential, histologically similar to the salivary gland counterpart, characterized by the presence of a biphasic cellular pattern of myoepithelial and epithelial cells (basaloid and ductal). The others terms for this tumor are carcinoma adenoids cysticum, adenocystic basal cell carcinoma, and cylindromatous carcinoma. Adenoid cystic carcinoma is equally distributed between the two breasts, and in about 50 percent of patients, the tumor is found in the subperiareolar region. The lesions may be painful or tender, and unexpectedly cystic. A discrete nodule is the most common presentation. True to the molecular signature of basal-like tumors, adenoid cystic carcinoma of the breast is often estrogen and progesterone receptors negative and does not express HER-2 (triple negative) (1, 2, 3, 4, 5, 6).

The authors report a case of adenoid cystic carcinoma, and review clinical features, pathological findings and diagnostic criteria of this rare tumor.

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CASE REPORT

A 50 years female patient sought the service of mastology referring a painless lump in her right breast. Clinical assessment identified a firm nodule in the upper-lateral quadrant of the right breast, described as a well-defined nodule in mammography (BIRADS 4B), which was submitted to the needle biopsy by ultrasonography. The histopathologic evaluation of the lesion showed a well differentiated malignant neoplasm with epithelial differentiation, constituted by two distinct cellular patterns, which were grouped in a tubular / trabecular pattern, and with a surrounding fibrous stroma. One of the cellular pattern was represented by basaloid cells in continuity with the fibrous stroma. The other pattern was represented by cuboidal to spindle shaped cells with eosinophilic cytoplasm and oval nucleus and organizing glandular structures. The microscopic examination of the sample corresponded to adenoid cystic carcinoma of the breast (Figure 1). Immunohistochemistry evaluation of the tumor revealed negative staining for estrogen and progesterone receptors and for HER-2. The patient underwent simple mastectomy with assessment of the sentinel axillary lymph node. At gross examination the lesion corresponded to a circumscribed, solid, gray nodule measuring 1.1 cm in diameter. The microscopic evaluation of lymph node did not identified metastases in the various serial histological sections analyzed.

DISCUSSION

Breast adenoid cystic carcinomas more frequently arise in females over 50 years, but occasional cases have been described in males. The age distribution ranges from 38 to 82 years. The tumor usually presents as a palpable, discrete, firm mass. Although calcifications develop in these tumors, few have been detected by mammography, and in some cases the mammogram was reportedly negative. Mammography of clinically palpable tumors reveals a well-defined lobulated mass or an ill-defined lesion. Pain or tenderness described in a minority of cases has not been particularly correlated with the finding of perineural invasion histologically. Skin dimpling, ulceration, or peau d'orange have been reported in patients with superficial or large lesions. Nipple discharge is rarely an initial symptom, despite the fact that adenoid cystic carcinoma occurs more commonly in the central or subareolar part of the breast (1, 2, 7, 8, 9).

The adenoid cystic carcinomas are usually solid, gray, circumscribed lesions, with occasional microcysts, and vary from 0.7cm to 12.0cm, with an average of 3.0cm. Despite their well-defined gross borders, about 50% of these lesions have an invasive growth pattern microscopically. Microcystic areas formed by the coalescent spaces in dilated glands are seen in about 25% of tumors. Perineural invasion and lymphatic tumor emboli are uncommon findings (2, 5).

Adenoid cystic carcinoma of the breast is very similar to that of salivary gland, and consists of a biphasic cellular pattern represented by proliferating glands (adenoid component) and stromal or basement membrane elements (cylindromatous component). These components are rarely distributed homogeneously in each case. Three basic architectural patterns are seen: cribriform, trabecular-insular, and solid. There are also two types of spaces, mostly seen with the cribriform pattern. The type referred to as pseudolumens is the result of intratumoral invaginations of the stroma. This type of space is of variable shape, mostly round, and contains myxoid acidic stromal mucosubstances or straps of collagen. In the smallest spaces, the content is constituted by small spherules or cylinders of hyaline material related immunohistochemically to basal lamina. The second type is less numerous, and usually

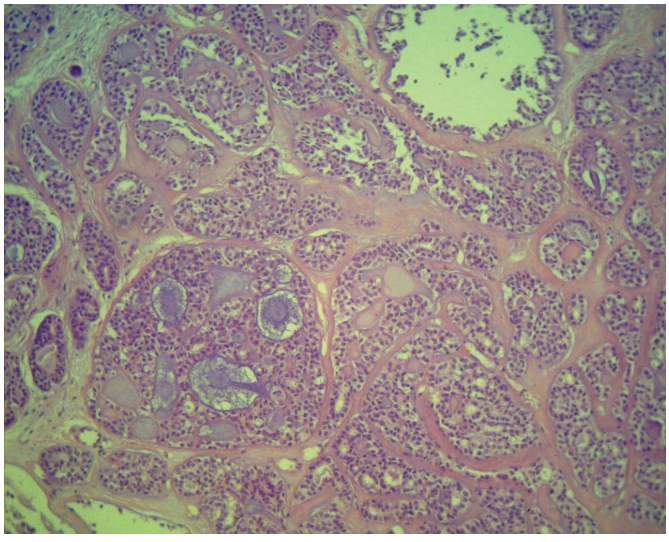


FIGURE 1 – Adenoid Cystic Carcinoma of the Breast: a biphasic malignant neoplasia constitutes by basaloid and cuboidal cells and a fibrous stroma, HE, 100x.

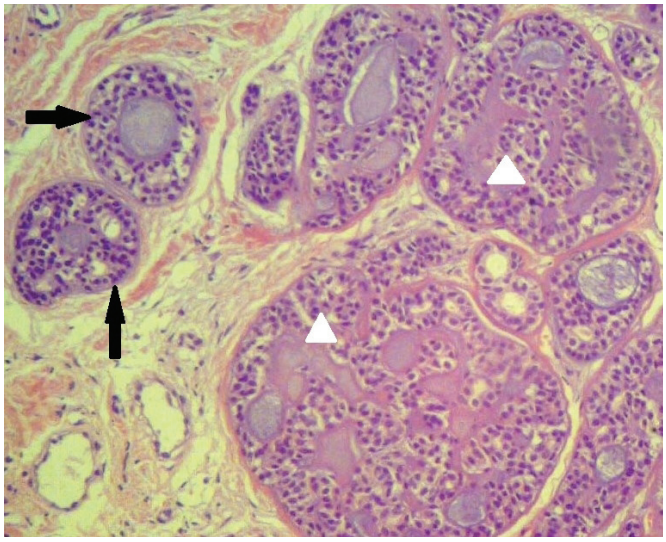


FIGURE 2 – Adenoid Cystic carcinoma of the Breast consists of a mixture of proliferating glands (adenoid component - arrowhead) and basement membrane elements (block arrows), HE, 200x.

composed of small lumens circumscribed for secretory glandular structures that contain eosinophilic granular secretion of neutral substances. Adenoid cystic carcinoma is composed by two different types of cells. One type of cell has scanty cytoplasm, a round to ovoid nucleus, and one to two nucleoli. This type of cell is also referred as basaloid cells, which comprise the majority of the lesion, and are found lining the cribriform stromal spaces. The second type of cells lines glandular lumens (ductal cells), which exhibited a cuboidal to spindle shape, and contains eosinophilic cytoplasm and round nuclei. These two cells types may be difficult to distinguish histologically. Alcian blue stains the contents of the pseudoglandular spaces, whereas the true lumina are PAS positive. The basaloid cells have myoepithelial features, and are immunopositive for actin, myosin, smooth muscle actin, S100, calponin and CK14. The cells that line the glandular lumens can have microvilli along the luminal margins (secretory type), or show abundant tonofilaments (adenosquamous type). The secretory type is positive for CK7, while adenosquamous cell are positive for CK7 and CK14. With occasional exceptions, adenoid cystic carcinoma of the breast is negative for estrogen and progestogen receptors, and for HER-2. The epithelial cells stain with antibodies to cytokeratin, such as Cam5.2, and 50% of the cases are c-kit positives. Some cases can show sebaceous, or adenomyoepitheliomatous, or syringomatous differentiation. Basement membrane material can be immunopositive for laminin and collagen IV. The tumoral stroma varies from tissue similar to that seen in normal breast to desmoplastic, myxoid, or even extensive adipose tissue. An *in situ* component is found in a minority of cases (1, 3, 4, 9, 10, 11, 12, 13, 14, 15).

Ro et al. suggested that adenoid cystic carcinomas may be stratified into three grades on the basis of the proportion of solid growth within the lesion (I: no solid elements, II < 30% solid; III > 30% solid). In high grade lesions, tumors cells are poorly differentiated, with sparse cytoplasm and large hyperchromatic nuclei. High grade lesions do not differ from lower-grade tumors in regard to patient age, laterality, duration prior to treatment, or hormone receptors (16). Adenoid cystic carcinoma is a low-grade malignant tumor that is generally treated by simple mastectomy. At present, no lymph node excision is indicated in adenoid cystic carcinoma. The tumor rarely spreads via the lymphatic system (1, 2, 7, 9, 14).

The main differential diagnosis is with invasive cribriform carcinoma of the breast, which is a tumor composed solely by epithelial cells, with a tubular elements associated to a stromal fibrous reaction, and it is typically positive for estrogen receptor. Adenoid cystic carcinoma must be distinghuished too from collagenous spherulosis, which is characterized by a hyperplastic epithelium that forms true glands and acellular sperules, creating an adenoid cystic structural arrangement. The lumens of the glandular spaces tend to have a more irregular shape than in adenoid cystic carcinoma. The attenuated myoepithelial cells are difficult to identify in hematoxylin-eosin sections, but can be highlighted by immunostains for actin and S-100 protein (1, 5, 7, 14, 15).

FINAL COMENTS

The authors reported the clinicopathologic features of an adenoid cystic carcinoma arising in the upper-lateral quadrant of the right breast in a 50 years female patient, measuring 1.1cm in diameter and showing negative imunoexpression for the hormonal receptors and HER-2. Adenoid cystic carcinoma of the breast is a rare tumor that is associated with a favorable survival, although the basal phenotype (standard triple negative) that is usually observed in more aggressive breast carcinomas.

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