Intra-cystic papillary carcinoma of the breast: Report of three cases

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Abstract

Intra-cystic papillary carcinoma of the breast is a rare malignant tumor. It represents less than 2% of all breast cancers, characterized by a slow growing with a good prognosis and low rate of local and distant recurrences.

Three cases were collected in Department of Pathology at JRA University Hospital. The aim was to report the epidemiological and histopathological features of the lesion through the review of literature. The patients, two women and one man, were respectively 63, 80 and 75 years old. Clinically, it manifested as a breast mass. The lesions were on the left side in 2 cases (women's cases). For the man's case, the lesion was on the right side and he was undergone for excisional biopsies. The histological diagnosis was intra-cystic papillary carcinoma.

Diagnosis of intra-cystic papillary carcinoma of the breast may be difficult at classical histological examination and identification of the myoepithelial cell layer by immunohistochemical study is helpful.

Key words: breast cancer, papillary carcinoma, myoepithelial cells

Introduction

Intracystic papillary carcinoma is a rare form of breast cancer [1, 2]. It accounts 0.5 to 2.4% of breast cancers in women and 5 to 7.5% in men [3]. It is the second type of breast cancer in men [4]. The clinical manifestation and the radiological aspects are nonspecific, and present differential diagnosis with other types of cancer [4]. Due to its rarity, there are limited data regarding the tumor and most studies come in the form of small case series. The aims of the present study were to report three cases, diagnosed in the Department of Pathology at JRA University Hospital Antananarivo, and to describe the epidemiological and histopathological features of the lesion with review of the literature.

Observations

Case n°1

A 63-year-old woman presented a left breast mass on the upper external quadrant. Mammography with ultrasound revealed a well-defined 5 cm mass and classified ACR 4. There was no associated axillary lymphadenopathy. A surgical excision of the external quadrant was performed.

Grossly, the specimen was 12 cm in diameter, surrounded by a skin flap. The excised tissue showed a friable, well circumscribed nodule measuring 5x4x3cm,.

On histological examination, the mass was composed by a tumor proliferation with papillary architecture within a cyst, composed by fibrovascular stroma, lined by cells with moderate cytonuclear atypia, often stratified. No infiltrating component was observed. The diagnosis was intracystic papillary carcinoma.

Case n°2:

It was an 80 years old woman with a left breast mass on the lower external quadrant. Mammography and ultrasound found a well-defined mass measuring 3 cm, classified ACR 3. There was no associated axillary lymphadenopathy. Excisional biopsy of the mass was performed. Grossly, the excised tissue was nodular, measuring 4 cm in diameter, polycystic with a nodule size 3x2x1.5 cm within a cyst. On histological examination, the nodule was an intracystic tumor proliferation composed by cells with moderate cytonuclear atypia, with papillary pattern, floating in the cavity, not exceeding the basal membrane of the cyst (Fig 1). It was surrounded by a thick fibrouscapsule. Thus, it was an intracystic papillary carcinoma.

Case n°3:

A 75-year-old man presented a tumor in the right breast. Clinical examination showed a mobile mass, subareolar. No radiological examination was performed. He was undergone for a surgical excision of the nodule.

Grossly, the specimen has 4 cm in diameter, contained a 2 cm of well-circumscribed nodule.

The histological features of the nodule were an intra-ductal proliferation of carcinomatous cells with moderate cytonuclear atypia, papillary architecture without myoepithelial cells, centered by fibrovascular stalks. No infiltrating component was observed. The diagnosis was intracystic papillary carcinoma.

Discussion:

Intra-cystic papillary carcinoma is defined as a solitary malignant breast tumor in situ, located in an encysted or dilated milk duct. In 1980, a classification system was developed and divided papillary carcinomas into two entities, invasive and non-invasive [5]. Carter et al subdivided non-invasive papillary carcinoma into two types. The diffuse form, a papillary variant of ductal carcinoma in situ and the localized form, intracystic (encysted) papillary carcinoma [4, 6]. The latter generally occurs in women after 40 years, more frequently in postmenopause with an average age varies from 55 to 67 years [7]. In the study of 917 patients made by Julia Grabowski et al [5], for example, papillary carcinoma is seen at any age and the average age of patients was 69.5 years. In men, intracystic papillary carcinoma also occurred in old man and the peak was found between 68 and 84 years age group [8].

The clinical and radiological manifestations of the pathology are not specific [4]. Clinically, it appears as a palpable, benign-like mass (in more than 80% of cases) with bloody nipple discharge (up to 22%) [5, 9]. In 50% of cases, it is in the retroareolar region according to Abderrahman EM et al [10]. For our female cases, the mass was on the external quadrant, it is in agreement with the location far from the nipple proposed by Darryl Carter et al [2]. But for the man’s case, it is under the nipple, this can be explained by the small size of the male breast tissue.

In our study, the nodules had 2 to 5 cm in diameter. According to the literature, the tumor size varies from 1 to 14 cm [10], it can be large when the cystic component is important. The presence of axillary lymphadenopathy is rare [11].

On ultrasound, the appearance of the lesion is nonspecific; it may be a pure cyst, a solid mass or mixed [4]. The mammographic appearance of intracystic papillary carcinoma is less specific. Small lesions are not often identified, while larger lesions may resemble any other well-circumscribed focal dense mass [12]. In our study, radiological examination (for two patients) revealed a well-limited nodule, classified respectively ACR 4 and ACR 3.

The diagnosis of intracystic papillary carcinoma based on histological examination. Biopsy samples must involve the fleshy and solid component. On a mastectomy piece, these are circumscribed, cystic and often necrotic lesions. Macroscopic examination finds a polylobed, friable and hemorrhagic formation within a cyst with a thick, fibrous wall. On microscopic examination, the tumor architecture is usually papillary with cribriform pattern surrounded by a thick fibrous capsule, lacking a layer of myoepithelial cells around the proliferation [5, 9, 10, 13]. The absence of a regular myoepithelial layer is the most important features to identify, which may be focal but not continuous, and the generally delicate, slender appearance of the fibrovascular papillary axes, which often consist of a few capillaries supported by a small amount of fibrous stroma [2].

The presence of a layer of myoepithelial cells at the periphery of areas of papillary carcinoma has historically been used to define carcinoma lesion in situ rather than invasive. Recent studies have shown that, unlike ductal carcinoma in situ of papillary architecture, intracystic papillary carcinoma does not appear to have a layer of myoepithelial cells surrounding the tumor nodules. The absence of myoepithelial cells suggests that intracystic papillary carcinoma may not be a lesion in situ but a localized, circumscribed and very low-grade form of invasive carcinoma or may be part of the intermediate progression between in situ and invasive lesions. It can potentially cause distant metastases [7, 14]. Therefore, the term encapsulated papillary carcinoma has been used when the myoepithelial layer is not identifiable [2, 5].

Histologically, there are three main subtypes of intracystic papillary carcinoma, the pure form, that associated with ductal carcinoma in situ, and that associated with invasive carcinoma [4, 6]. An infiltrating component, usually minimal, may develop and penetrate the wall of the cyst [11]. All our cases were pure form.

There are no well-defined views on the management of the intra-cystic papillary carcinoma However, several studies report an excellent prognosis with conservative surgery without lymph node dissection [1]. Lefkowitz reports a 10-year survival rate of 91%.

Conclusion

Intra-cystic papillary carcinoma is a rare breast tumor, usually occurs in postmenopausal women but can occur at any age and in men. The diagnosis is not always suspected before surgery. The clinical and radiological features are non-specific. The positive diagnosis is histological. Treatment with conservative surgery gives a good prognosis.

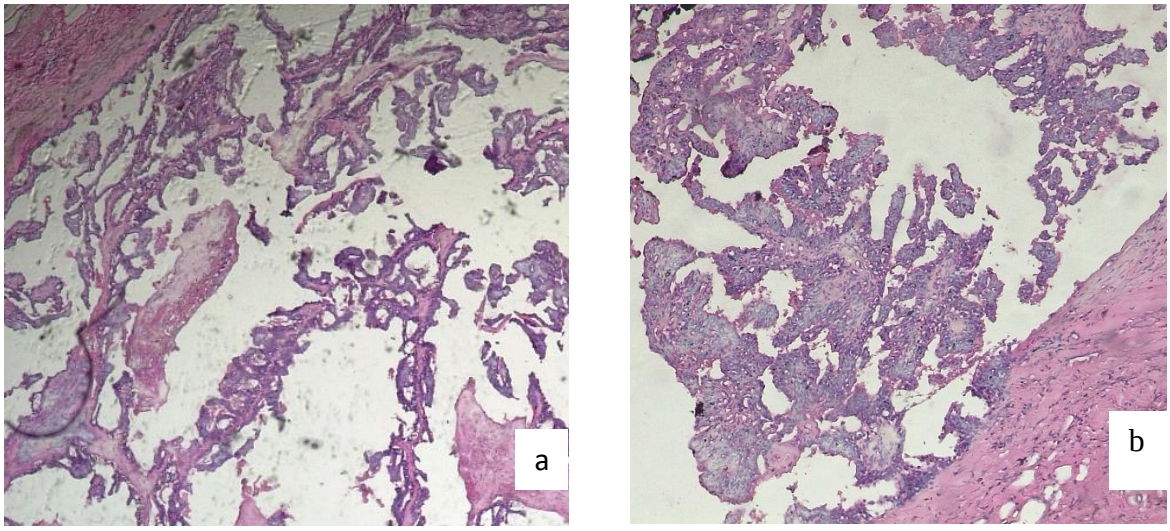

Figure 1: Breast mass. Intracystic papillary carcinoma composed by cells with moderate cytonuclear atypia, with papillary pattern within a cyst, surrounded by a thick fibrouscapsule. HE x 40 (a), HE x100 (b).
Source: Department of Pathology at JRA University Hospital, Antananarivo Madagascar.

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