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Intracystic papillary carcinoma of the breast: Report case and literature review

Benmouna Imane *, Mouimen Soukaina, Slaoui Aziz, Pr Baidada Aziz

Department of Gynecology and Obstetrics and Endoscopy, Maternity Souissi Hospital, IbnSina University Hospital, Mohammed V University, Rabat, Morocco.

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Abstract

Intracystic papillary carcinoma (IPC) of the breast is a rare malignant tumour, found mainly but not exclusively in elderly women. IPC may be asymptomatic or presents with a palpable mass or blood-stained nipple discharge. Radiologic manifestations of IPC are not specific. On ultrasonography, it can be a pure cyst, a mixed image, or a solid mass. Histologic features of the tumor include cellular proliferations surrounding fibrovascular cores, with or without invasion. The mainstay of treatment is breast-conserving surgery or mastectomy. Sentinel node biopsy could be considered in invasive cases. Adjuvant radiotherapy and/or endocrine therapy is considered in appropriate cases. Through the observation of a 58-year-old patient, we report the epidemiological, clinical and radiological data of papillary breast carcinoma.

Keywords: Intracystic Papillary Carcinoma; Ultrasound; Mammography

1 Introduction

Intracystic papillary carcinoma of the breast is a very rare pathology, representing between 0.5 and 2.4% of cancers of breast cancer in women, the prevalence of both invasive and in situ papillary carcinoma seems to be greater in older postmenopausal women and, in relative terms, in males. Distinction of invasive papillary carcinoma from non-invasive forms is critical, as each entity carries a unique prognosis [1].

Patients with IPC may present with a palpable mass, bloody nipple discharge, or a radiographic abnormality.

Histologically, the tumor is encysted within a dilated duct with arborization of the fibrovascular stroma and contains nodules of papillary carcinoma surrounded by a thick fibrous capsule [2].

The recent use of immunohistochemical identification of myoepithelial cells layer (MEC) of the cysts seems to enhance pathologic diagnosis performance but its influence on prognosis is to be determined.

Therapeutic management of IPC is also still controversial. Endocrine therapy and radiation are used by many centers but evidence of their role in prognosis improvement is still lacking.

*Corresponding author: Benmouna Imane
Department of Gynecology and Obstetrics and Endoscopy, Maternity Souissi Hospital, Ibn Sina University Hospital, Mohammed V University, Rabat, Morocco.

2 Case observation

A 58-year-old lady presented with a 2-week history of right breast lump and no pain or nipple discharge. Her medical history was unremarkable,Physical examination revealed a 2.5 cm, mobile mass in the upper outer quadrant of the right breast.nonpainful, and without skin changes.There was no evidence of axillary lymphadenopathy.

The mammogram showed dense, round, circumscribed mass without calcification, classified as a Bi-RADS4(Figure 1). Ultrasonography showed a complex mass with cystic and solid components measuring 2, 41×1.78 cm (Figure 2-3). The mass also showed some posterior acoustic enhancement. US of axilla showed no suspicious lymphadenopathy. Color Doppler of ultrasonography showed a surrounding vascular pattern.

US-guided core biopsy showed dyscohesive cells which appear to be epithelial in origin with mild nuclear pleomorphism, highly suggestive of malignancy. The patient underwent right mastectomy and axillary dissection. The histopathologic diagnosis was intracystic papillary carcinoma, node negative.

She was referred to oncologist for radiotherapy and hormonal treatment as the tumour cells were 100% oestrogen and progesterone receptor (ER/PR) positive. There was no recurrence at 13 months of follow-up.

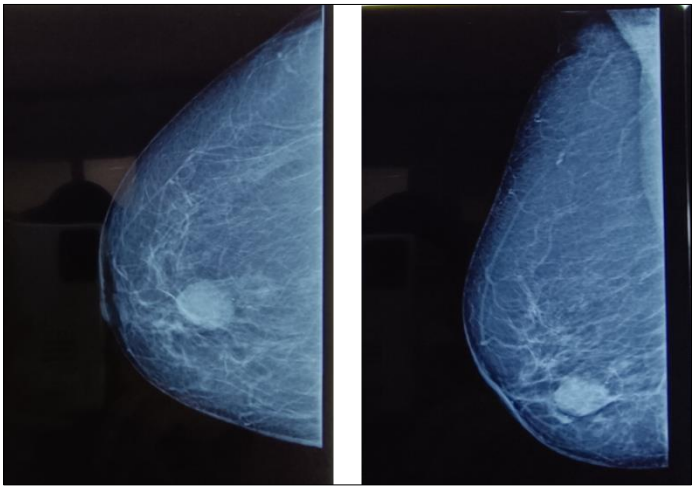

Figure 1 Mammography (a, b). Mammography showed dense, round, circumscribed mass without calcification

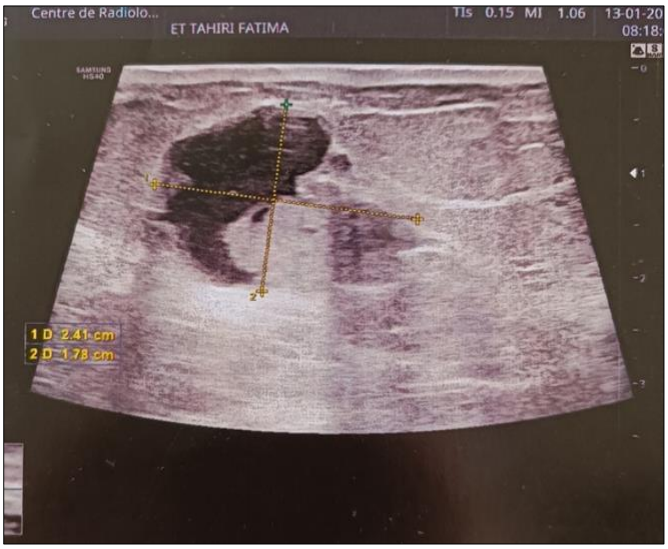

Figure 2 Ultrasonography showed complex mass with cystic and nodular solid, components. Posterior acoustic enhancement is also seen

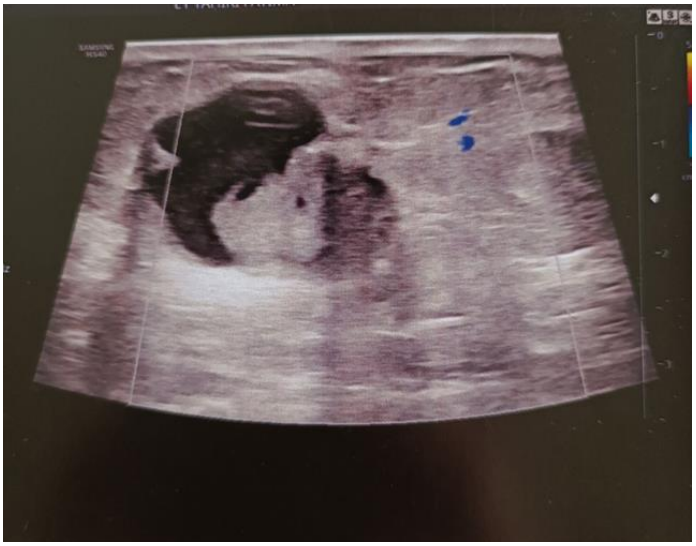

Figure 3 Colour Doppler of ultrasonography showed a surrounding vascular pattern

3 Discussion

These lesions most commonly occur in older women [3], have been termed intracystic or ‘‘encysted’’ papillary carcinomas, IPC is not an entity unique to women, occasionally being found in men [4].

Papillary carcinomas are classified histologically into intraductal and intracystic papillary carcinoma. Intracystic papillary carcinomas (IPCs) are further divided into pure form or associated to a ductal carcinoma in situ (DCIS) or invasive carcinoma [4,5].

In about 50% of the cases it is centrally located and more precisely in the retroareolar region. [6] It may manifest with no symptoms, as a slowly enlarging palpable mass, [7] or as bloody nipple discharge (about 22%–34% of cases). [5]it can be asymptomatic and revealed by systematic mammography. Axillaries nodes are infrequent [5,8].

On mammography, papillary intracystic carcinoma usually appears as a well-circumscribed ovarian or lobular mass. The contours are usually well circumscribed but sometimes they can be locally masked or indistinct, [5,8] spiculated contours are rare (4). Several diagnoses can be evoked in front of this mass: hematoma, infiltrating ductal carcinoma, mucinous carcinoma, medull. Cysts and adenofibromas are less frequent at this age and remain diagnoses of elimination [9]

Ultrasonography usually shows solid hypoechoic mass, or a complex mass with cystic and nodular solid components with posterior acoustic enhancement [2,5]. Color Doppler eliminates the diagnosis of a remodeled cyst with thick contents and demonstrate the intramural blood flow within the solid component of the mass [10]. These papillary lesions tend to bleed spontaneously, thus exfoliating the echogenic content and the declipped cellular debris. [11] The presence of a large solid portion and signs of spontaneous intracystic bleeding is more suggestive of papillary carcinomas than of benign papillomas. [11] In rare cases, a cystic carcinoma might have no solid component [12].

Magnetic Resonance imaging of the IPC is sensitive but not specific in detecting papillary tumors. It shows mural nodules and internal septa or might show a multicystic appearance.

Preoperative ultrasound-guided Fine Needle Aspiration yields a bloody or old-blood-colored liquid, and the cyst recurs rapidly after Aspiration [8], although the bloody aspirate is not pathoganomonic for IPC. Cytological examination should be interpreted with caution because of the frequency of false negative results due to necrotic materials, degenerative changes in the diagnostic cells and abundant obscuring blood in the cystic lesion [8,13]. Core biopsy is a useful tool for diagnosis of IPC, although it is important to keep in mind that differentiation between in situ and invasive PC is difficult because the centre of the lesion is often targeted, and invasion is often found at the periphery of the tumour. Therefore, excisional biopsy is often performed when IPC is suggested [4,5,14].

Macroscopic examination reveals a a cyst surrounded by a fibrous wall with a friable, hemorrhagic tissue formation [15]. Histologic features include arborization of fibrovascular stroma covered by a single or multiple layers of cells and a lack of myoepithelial cells [2,16,17]. Intracystic papillary carcinoma can show four cellular patterns: cribriform, compact columnar epithelial, stratified spindle cell, or a transitional cell form resembling urothelium, or a combination of two or more of these patterns may be seen . IPC may be associated with foci of DCIS or invasive cancer, and necrosis is often a prominent feature when an associated invasive component is present [18].

There are no clear guidelines for the management of IPC. [19] If it is a case of pure IPC, Several studies have shown that prognosis is excellent and recurrence rates low, regardless of the intervention. [4,20,21] Carter et al. [22] in a series of 7 cases of pure IPC treated by complete local resection did not observe any local recurrence after a 7-year follow-up.

However, this will change depending on whether DCIS exists outside the main tumor mass or an invasive component is present. Most early researchers failed to distinguish between these different patient groups, and the overall impression was that IPC had an unfavorable prognosis and should be treated with radical mastectomy [23].

Since less than one percent of IPC without ductal carcinoma in-situ (DCIS) is associated with lymph node metastasis, Harris recommends treating pure IPC with wide local excision without axillary lymph node dissection. [20,23]

However, Axillary lymph node dissection or sentinal node biopsy is often performed in patients in whom invasion is likely [14, 17, 22, 24].

The need for radiotherapy and chemotherapy in IPC, IPC with DCIS, and IPC with invasion also remains unclear [16,23].

Fayanju et al. [21] Concluded that the most important factor determining the use of radiation and endocrine therapies is associated pathology (DCIS or microinvasion) and patients with pure IPC were less likely to undergo radiation and endocrine therapies.

Even if IPC is associated with breast DCIS or invasion, the prognosis still excellent when treatment decisions are tailored to associated pathology. [21]Lefkowitz [25]reports a 10-year disease-free survival rate of 91%.

4 Conclusion

IPC is a rare tumour with an excellent prognosis. Suspected at sonography in front of cyst with internal solid component, it will be confirmed by histopathology and immunohistochemical study after surgical excision. An appreciation of associated pathology with IPC may be critical in surgical decision-making.

Compliance with ethical standards

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Disclosure of conflict of interest

The authors declare that they have no conflict of interest in relation to this work.

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