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Intracystic Papillary Carcinoma of the Breast with Associated Invasive Carcinoma: A Rare Case Report.

Hema Kumar, Divya Achutha Ail*, and Geetha Prakash.

Department of Pathology, Meenakshi Medical College and Research Institute, Enathur, Kanchipuram - 631 552, Tamilnadu, India.

Case Report

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***For Correspondence**

Department of Pathology, Meenakshi Medical College and Research Institute, Enathur, Kanchipuram - 631 552, Tamilnadu, India.
Mobile: +91 9884995621

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ABSTRACT

Intracystic papillary carcinoma (IPC) of the breast is an uncommon malignant breast neoplasm representing approximately 0.5% to 2% of all breast cancers and typically occurs in post-menopausal women. We report this case of intracystic papillary carcinoma solid variant associated with foci of invasive ductal carcinoma in a 65 year old female who presented with a painless lump in the left breast associated with axillary lymphadenopathy. Pure intracystic papillary carcinomas have a slow growth rate and excellent prognosis, but the recognition of a coexistent lesion such as ductal carcinoma in situ or invasive ductal carcinoma is very important for which careful pathological examination is essential.

INTRODUCTION

Intracystic papillary carcinoma (IPC) of the breast is an uncommon malignant breast neoplasm. It represents approximately 0.5% to 2% of all breast cancers and typically occurs in post-menopausal women [1]. It is characterised by a more benign behaviour and a subsequent higher survival rate [2]. Pure intracystic papillary carcinomas have a slow growth rate and an excellent prognosis with a ten year survival approaching 100% [3], but the recognition of a coexistent lesion such as a ductal carcinoma in situ or invasive carcinoma has an adverse effect on the prognosis. We present here a case of a 65year old female who presented with a slow growing mass in the left breast. Examination revealed a painless well circumscribed mass hard in consistency. Fine needle aspiration cytology, mammography and ultrasound proved inconclusive. Based on the preoperative clinical identification of a left axillary lymphadenopathy, the patient underwent a modified radical mastectomy with a level 1 axillary clearance. Histology revealed an intracystic papillary carcinoma solid variant with an associated focus of invasive ductal carcinoma.

Case Report

A 65 year old female presented with a mass in the upper inner quadrant of the left breast. Examination found a painless well circumscribed mass hard in consistency measuring 4cm, with retraction of the nipple areola and a solitary palpable axillary node measuring 2.5x1cm. The mass was lobular in structure without calcifications. On sonography the mass was heterogeneous with solid and pinpoint cystic areas. A fine needle aspiration biopsy of the mass revealed a diagnosis of suspected invasive carcinoma. A modified radical mastectomy with a level 1 axillary clearance was performed. There was a 4cm sized tumour, which was solid grey white irregular with small cystic areas.

On microscopic examination, the tumour was composed of malignant ductal epithelial cells with vesicular nuclei arranged in papillary fronds projecting into large cystic spaces alongside solid nests of

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cells with similar morphology (Figure 1a, 1b). Other foci showed malignant cells with a higher nuclear grade arranged in acinar and comedo patterns, focal areas of lobular differentiation seen (Figure 2a, 2b). Ducts adjoining the papillary foci showed marked periductular hyalinisation. 6 out of the 12 nodes identified showed similar tumour deposits. The nipple areola and the surgical resected margins showed tumour infiltration. Based on the above findings a final diagnosis of intracystic papillary solid carcinoma associated with a infiltrating ductal carcinoma was made. Immunohistochemistry revealed absence of myoepithelial cell markers (MEC) such as smooth muscle actin and P63 and positivity for estrogen and preogesterone receptors with negativity for Her2/neu.

Following the surgery chemotherapy or radiation was not given and there was no recurrence on 11 months of follow up.

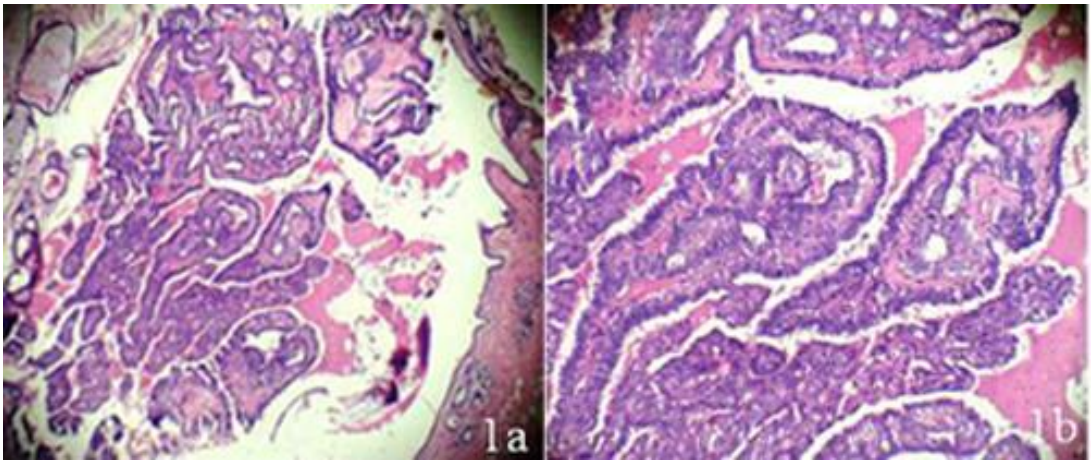

Figure 1a: Malignant ductal epithelial cells arranged in papillary fronds projecting into large cystic spaces [Haematoxylin and Eosin (H and E) x40].
Figure 1b: Malignant epithelial cells in papillary fronds showing nuclear clearing (H and E X100).

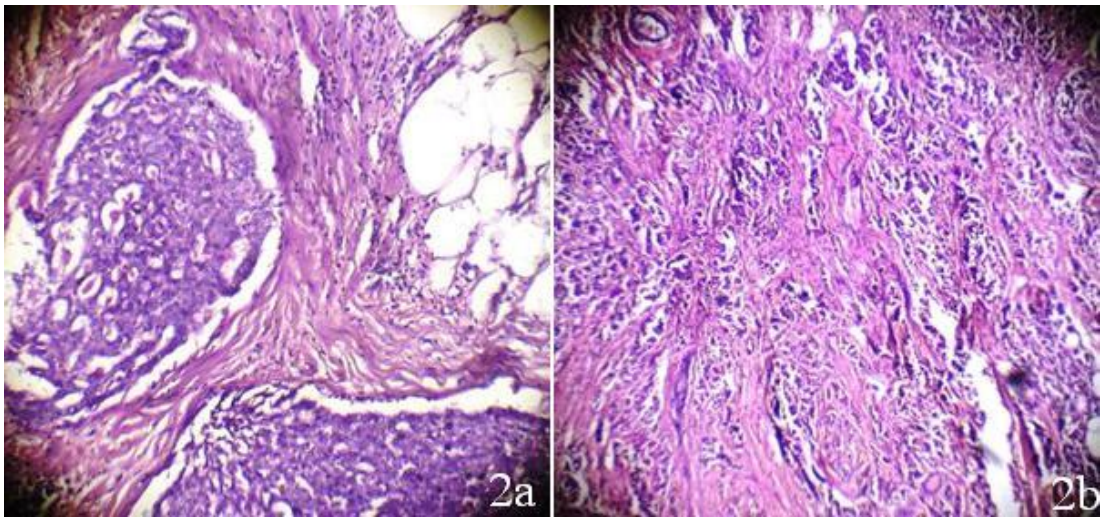

Figure 2a and 2b: Foci of malignant ductal epithelial cells arranged in cribriform and India file pattern respectively (H and E X 100).

DISCUSSION

Papillary carcinomas constitute less than 2% of breast carcinoma. It is mostly an in situ carcinoma, namely "insitu papillary carcinoma", but in a small group stromal invasion occurs, in which case the lesion is called an "invasive papillary carcinoma". Histologically whether insitu or invasive papillary carcinoma is further classified as "intraductal papillary carcinoma" when the duct simply expands to accommodate the proliferating lesion, "intracystic papillary carcinoma" (IPC) [4] if it becomes cystically dilated and "solid papillary carcinoma" when there are nodules formed by proliferating epithelial cells [4].

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Intracystic papillary carcinoma is more frequently found among the post-menopausal women with an average age between 55 and 67 years old [5]. Clinically it may be asymptomatic, or present as a slowly enlarging palpable mass or as a bloody nipple discharge. Retraction of the nipple and skin may be associated clinical finding [3].

At mammography, intracystic papillary carcinoma appears as a round, oval or lobulated opacity. The margins of the mass are usually circumscribed, but may be obscured or indistinct at places testifying inflammation or invasion. The differential diagnosis on mammography in the age group of these tumors includes hematoma, invasive ductal carcinoma, colloid or medullary carcinoma, benign cyst or adenofibroma [5].

At sonography, intracystic papillary carcinoma appears as single or predominantly cystic masses, with or without septa and with solid papillary mass projecting into the cystic lumen from the inner wall [1]. Although some radiologic features, such as posterior acoustic enhancement and associated microcalcifications, are more frequently associated with malignancy, the radiologic appearance cannot accurately predict the behaviour of papillary lesions, and histologic evaluation is necessary [4].

Fine needle aspiration and core needle biopsy may be unable to differentiate between insitu and invasive lesions as invasion is often identified at the periphery of the tumor. Therefore surgical excision is performed for adequate histologic diagnosis and treatment [3].

Intracystic papillary carcinoma is accepted as a borderline lesion in progression from insitu to invasive carcinoma because histologically there is scant or no MEC layer a situation similar to invasive carcinoma [5]. It is histologically characterised by one or several nodules of papillary carcinoma surrounded by a fibrous capsule, it may have various cellular patterns (cribriform, stratified spindle cell, compact columnar epithelial or transitional cell form) or a combination of patterns and shows typical histologic findings, which are low or intermediate nuclear grade, absence of necrosis. High nuclear grade and the presence of necrosis seem to be indicator of more aggressive tumors [3].

A minority of encapsulated papillary carcinomas may be associated with a component of invasive carcinoma. The invasive component is characterised by an infiltrative appearance with extension beyond the fibrous capsule of the lesion. Invasive areas in general do not display papillary features but rather exhibit the morphology of an invasive ductal carcinoma not otherwise specified [6].

Solorzano et al. suggested that the standard treatment of IPC should be based on the associated pathology [7]. There are no evidence based guidelines for treatment of IPC. There is no randomized controlled trials comparing breast conserving surgery to mastectomy, however many case reports and retrospective studies show excellent prognosis with conservative surgery [5, 7, 8]. In cases associated with invasive carcinoma, the prognosis will depend on the invasive component of the tumor [9].

There has been no clear indication for adjuvant radiation and endocrine therapy, in their series of 40 patients associated with pure IPC, IPC associated with ductal carcinoma in situ and IPC associated with invasion, Solorzano et al reported that the use of radiation did not influence recurrence or survival. [7] In a similar study, Fayanju et al found that the most important factor determining the use of radiation and endocrine therapies is associated pathology and patients with pure IPC were less likely to undergo radiation and endocrine therapies [8,9].

CONCLUSION

Intracystic papillary carcinoma is a rare breast malignancy, with an excellent prognosis in its pure form. These tumors often demonstrate a paucity of myoepithelial layer on immunohistochemistry or electron microscopy, raising the concern of a pushing border invasive carcinoma. The presence of an associated invasive component has a negative impact on the prognosis. The mainstay of treatment is surgical resection, with adjuvant therapy if associated with insitu or invasive carcinoma.

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