

Encapsulated papillary carcinoma of the breast: A rare entity

Niharika Shah, Santosh Upadhyaya Kafle, Punam Paudyal

ABSTRACT

Introduction: Encapsulated papillary carcinoma (EPC) is a rare breast malignancy with a slow growth rate and excellent prognosis. **Case Series:** There were two cases of EPC, one of them was a 60-year-old female presenting with two lumps in the right breast at 3 o'clock and 9 o'clock positions. A right modified radical mastectomy with axillary dissection was done subsequently. On histopathology EPC was diagnosed. The other case was of a 45-year-old female presenting with bloody nipple discharge and a 2-cm subareolar lump. Histopathology revealed features of EPC with invasion into the fibrous capsule. In both cases, there was no associated ductal carcinoma in situ (DCIS) in the surrounding tissue and lymph nodes were not involved. **Conclusion:** Encapsulated papillary carcinoma is a rare breast malignancy with excellent prognosis. In the absence of associated DCIS in the surrounding tissue or areas of infiltrating carcinoma, it has a very favorable prognosis with adequate local therapy alone.

Niharika Shah¹, Santosh Upadhyaya Kafle², Punam Paudyal³

Affiliations: ¹MD Pathology, Assistant Professor, Department of Pathology, B.P. Koirala Institute of Health Sciences, Dharan, Nepal; ²MD Pathology, Fellowship Dermatopathology, Associate Professor, Department of Pathology, Birat Medical College, Biratnagar, Nepal; ³MD Pathology, Associate Professor, Department of Pathology, B.P. Koirala Institute of Health Sciences, Dharan, Nepal.

Corresponding Author: Niharika Shah, Assistant Professor, Department of Pathology, BPKIHS, Ghopa Camp, Dharan, Nepal, 56700; Email: niharika.shah@bпкиhs.edu

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INTRODUCTION

Encapsulated papillary carcinoma (EPC) of the breast, also known as intracystic or encysted papillary carcinoma represents approximately 0.5–2% [1] of all breast cancers and occurs typically in postmenopausal women. Encapsulated papillary carcinoma (EPC) is characterized by papillary carcinoma within a well-circumscribed cystic or distended duct. It has only been recognized since some time that these lesions lack myoepithelial cells at their periphery. Thus some authors are even of the opinion that EPCs are, in fact, invasive carcinomas with an expansile growth pattern which was supported by a study that reported lung metastases developing in a patient after a diagnosis of EPC, and another that reported two cases with axillary lymph node micrometastases [1].

Encapsulated papillary carcinoma mostly presents as a circumscribed round mass with or without nipple discharge making them indistinguishable from those of benign papillary lesions. No specific clinical or imaging features distinguish EPCs from other papillary lesions. Nevertheless, EPCs tend to be larger at presentation [2].

CASE SERIES

Case 1

A 60-year-old female presenting with two lumps in the right breast at 3 o'clock and 9 o'clock positions. Radiological data suggested a cystic breast lesion with a BI-RADS (The Breast Imaging Reporting and Data System) category IV (suspicious abnormality) on mammography [3].

The patient had also undergone a FNAC before the surgery with a cytological diagnosis of intracystic neoplasia. A right modified radical mastectomy with axillary dissection was done subsequently and the specimen was received along with axillary tail with overlying skin flap. The nipple and areola were unremarkable and on serial sectioning two nodular masses were identified each measuring 2.5x2x1 cm and 0.7x0.5x0.3 cm. On histopathological examination, a well-circumscribed tumor surrounded by a fibrous capsule was identified. These tumor cells were arranged in papillary fragments, some exhibiting a cribriform pattern with overlying fibrovascular stalks. Myoepithelial cell component were not seen both within and at the periphery of the tumor both on H&E as well as with myoepithelial cell markers (SMA and S-100) (Figures 1 and 2). A diagnosis of EPC was made. The lymph nodes were free of metastatic tumor deposits.

Case 2

A 45-year-old female presenting with bloody nipple discharge and a sub-areolar lump in the right breast which on physical examination revealed a lump about 2 cm in size. An excision biopsy was done along with a single axillary lymph node. The specimen consisted of breast tissue which on serial sectioning revealed a lesion measuring 1.8x1 cm. The lesion was well capsulated, solid-cystic with solid area revealing papillary excrescences. Histopathological examination showed multiple cysts embedded in fibrous stroma. The cyst contained cells arranged predominantly in a papillary pattern and some in cribriform pattern as well. Myoepithelial cells were inconspicuous to absent on hematoxylin and eosin stain as well as with a myoepithelial cell marker (S-100). A diagnosis of EPC with invasion into the fibrous capsule was made. (Figure 3) The separately sent lymph node showed features of reactive lymphadenitis.

DISCUSSION

Myoepithelial cells are lacking in EPCs both within the fibrovascular cores and at the periphery of the lesion. This absence of a myoepithelial cell layer, both on hematoxylin and eosin stained sections and immunohistochemistry for a variety of myoepithelial cell markers, is contrary to our current understanding of such lesion [2]. This has been appreciated and only recently been documented in

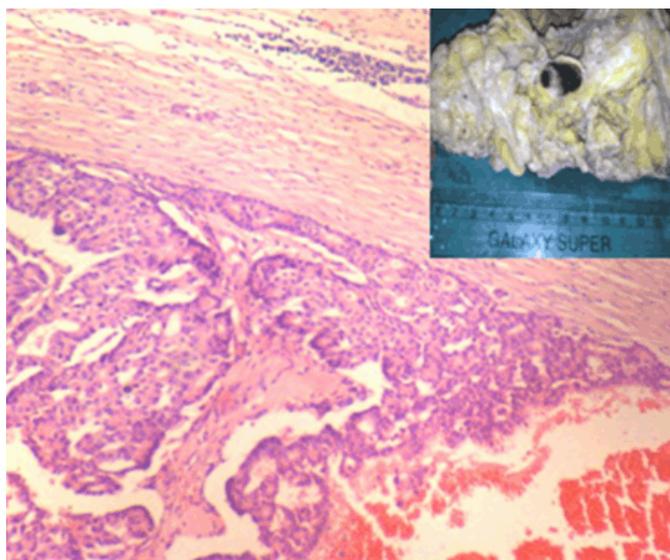


Figure 1: Encapsulated papillary carcinoma, Case 1, at low magnification revealing the papillary fronds lined by a monotonous epithelial proliferation (H&E stain, x100). (Inset: A well-capsulated cystic lesion was identified on gross examination).

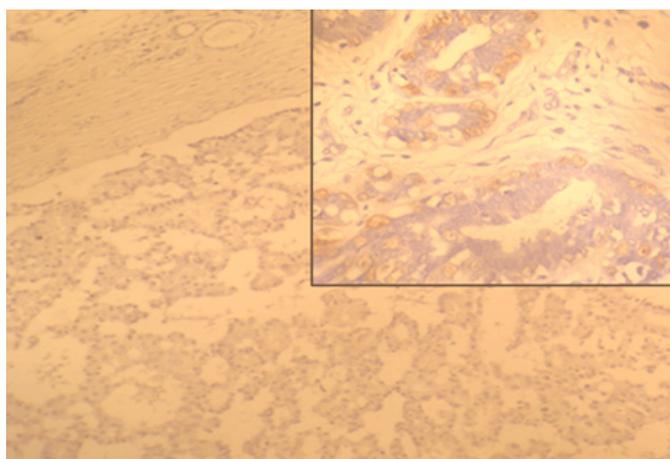


Figure 2: Myoepithelial cells are absent within and at the periphery of the tumor, as seen by absence of reactivity with S-100. (x100). (Inset: S-100 positive myoepithelial cells in normal breast tissue from the same case x200).

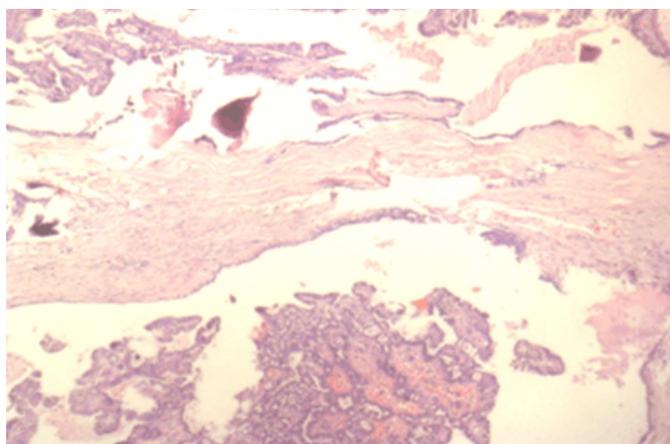


Figure 3: Focus of invasion in low power just adjacent to the encapsulated papillary carcinoma in Case 2 (H&E stain, x100).

literature, but it makes it more possible that EPCs may, in fact, represent a minimally invasive, low grade or indolent form of invasive carcinoma than an in situ lesion. While some are even of the opinion that these lesions may be a form of carcinoma ‘in transition’ between in situ and invasive carcinoma [2]. A diagnosis of frank invasive carcinoma should only be signed out when neoplastic epithelial elements are seen beyond the fibrous capsule of EPCs. In many instances, neoplastic epithelial cells are entrapped within the fibrous capsule and are even displaced into the biopsy site, mostly following needle core biopsy procedures of papillary lesions (and since immunohistochemical studies for myoepithelial cells will not be helpful in this setting, this is a differential that is best determined on histological examination) [2].

In a study conducted by Collins et al., they were unable to identify a delimiting layer of myoepithelial cell (MEC) layer at the periphery of any of 22 lesions classified as EPC using any of five sensitive MEC markers [smooth muscle myosin heavy chain (SMMHC), calponin, p63, CD10, and cytokeratins 5/6 (CK5/6)]. Perhaps these findings are an indicator that at least some or most lesions classified as EPC entirely on histologic grounds represent encapsulated nodules of invasive papillary carcinoma. Thus keeping these observations in mind, the term ‘encapsulated’ papillary carcinoma is preferable to intracystic papillary carcinoma for circumscribed nodules of papillary carcinoma surrounded by a fibrous capsule and in which a peripheral delimiting layer of MEC is not demonstrable. Regardless of all these observations. However, it is important to emphasize that no matter if these lesions are truly in situ or invasive carcinomas, available outcome data indicate that they have an excellent prognosis with adequate local therapy alone [4–8].

In one of our cases, a foci of true invasion was seen in the surrounding breast tissue, however, areas of ductal carcinoma in situ (DCIS) were not seen in both the cases and lymph nodes were negative for metastatic tumor deposits. According to the World Health Classification of tumors of the breast, the presence of associated DCIS in the adjacent breast tissue confers a higher rate of local recurrence. Thus, it is essential for treatment and assessment of risk for local recurrence that EPCs have complete surgical excision with extensive sampling of the lesion and surrounding breast tissue. Staging of EPCs, however has become controversial. If there is a component of invasive carcinoma associated with the EPC, the tumor should be staged according to the size of the invasive carcinoma. There is no universal agreement as of now on how to stage EPCs. When conventional invasive carcinoma is not present, the consensus of the WHO working group was that such lesions should be staged and managed as Tis disease [2].

CONCLUSION

It would thus be best to conclude that encapsulated papillary carcinoma (EPC) lacking myoepithelial cells are a special type of invasive breast carcinoma with favorable prognosis, whereas EPC showing an intact peripheral layer of myoepithelial cells should be regarded as in situ carcinomas. The term invasive papillary carcinoma should be abolished, and papillary carcinoma with coexisting conventional carcinoma should be named according to the non-papillary component. Encapsulated papillary carcinoma can be treated with adequate local therapy alone with or without hormonal therapy, as indicated in certain cases. The approach to lymph node sampling should be the same as for conventional ductal carcinoma in situ (DCIS) [9].

Author Contributions

Niharika Shah – Substantial contributions to conception and design, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Santosh Upadhyaya Kafle – Substantial contributions to conception and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Punam Paudyal – Substantial contributions to conception and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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