

Intra-cystic (Encapsulated) Papillary Carcinoma of Breast: A Case Report & Review of Literature

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1. Abstract

We are reporting a case of 62-year-old lady who presented with nipple discharge without palpable lesion and after radiographic and pathologic investigations, diagnosed as intracystic papillary carcinoma of breast. They account for less than 3% of all breast tumors and comprises a wide range of proliferative breast diseases. It is a difficult task to diagnose encapsulated papillary carcinoma. Most of the lesions present asymptotically with or without palpable lesion or nipple discharge. It is a tough job for pathologist to differentiate between invasive and non-invasive lesion. Non-invasive lesion without axillary lymphadenopathy has excellent prognosis in comparison to invasive lesion. Usually, adjuvant hormonal therapy is useful as these tumors express estrogen and progesterone receptors. A multidisciplinary approach of oncologist, pathologist and radiologist is required in diagnosing and treating encapsulated papillary carcinoma of breast.

2. Introduction

Papillary Breast Lesions (PBL) are considered as rarely encountered phenomenon of all breast lesions [1]. They express less than 2% of all breast cancers and less than 10% of all benign lesions [2]. They account for less than 3% of all breast tumors and comprises a wide range of proliferative breast diseases [3, 4]. These lesions pose a diagnostic challenge to the treating team due to their special characteristics. Most of the lesions manifest as intraductal lesions however papillary features have been noted in some of the invasive cancers [5]. PBL represents both, benign and malignant lesions which includes intraductal papilloma (IDP), IDP with atypia or ductal carcinoma in situ (DCIS), papillary DCIS, encapsulat-

ed papillary carcinoma (EPC), solid papillary carcinoma (SPC), invasive papillary carcinoma (IPC) and invasive micropapillary carcinoma (IMPC) [6]. Benign intraductal papillomas are the most common PBLs with other names as central or solitary papillomas [7]. It is very important to differentiate and diagnose benign and malignant papillary lesions as their management differs [8]. Papillary lesions are present in atypical ductal hyperplasia and DCIS lesions [9]. IPC and IMPC are very rare entities. Malignant lesions are usually low nuclear grade and manifest in an in-situ manner [10]. Most of the patients are asymptomatic and it's difficult to detect and diagnose PBL in these patients [11]. Most commonly they present with palpable breast lesion with or without nipple discharge. We are reporting a case of 62-year-old lady who presented with nipple discharge without palpable lesion and after radiographic and pathologic investigations, diagnosed as intracystic papillary carcinoma of breast.

3. Case Report

A 62-year-old lady with Eastern Co-operative Oncology Group Performance Status I (ECOG PS- I) without any co-morbidity presented with serous nipple discharge from the right breast for the last 6 months. Patient underwent evaluation at a general surgeon's clinic. She had been advised bilateral sono-mammography which was suggestive of a breast lesion of size 1x1cm located at 5 o'clock position at lower and inner quadrant of right breast of BIRADS III grade with another lesion of size 0.4x0.5cm at retro-areolar location with BIRADS II grade. The treating surgeon decided to go ahead with no active surgical intervention with close follow up. She had been called after 3 months for follow up evaluation.

Nipple discharge was persistent. She had been advised ultrasound guided core needle biopsy and which was suggestive of papillary carcinoma of breast. In view of carcinoma of breast, PET (Positron Emission Tomography) had been advised. PET was suggestive of same breast lesions as described in sono-mammography without any FDG avid uptake in the breast lesion and no lymphadenopathy in the ipsilateral axilla with no distant metastasis (Figure 1). With these reports she had been referred to our clinic.

On clinical examination, there was no palpable breast lesion or axillary lymph nodes except serous nipple discharge on squeezing the nipple. Contralateral breast and axilla were normal and rest of the systemic examination was unremarkable. Case was discussed in our institutional multidisciplinary tumor board and board decided to go ahead with Breast Conserving Surgery with sentinel axillary lymph node biopsy. Patient and her relatives were explained about the pros and cons of Modified Radical Mastectomy (MRM) versus Breast Conserving Surgery (BCS). They opted for MRM considering chances of recurrences in future. Patient underwent MRM and discharged on 1st postoperative day. Final histopathology report showed a circumscribed nodule with fibrous capsule which was suggestive of intra-cystic papillary carcinoma of breast without invasion with tumor size measuring 1.5x1cm (Figure 2). Total 12 lymph nodes were retrieved and all were free from metastasis. Immunohistochemistry was suggestive of estrogen and progesterone receptor positive with HER2Neu receptor negative. As per AJCC 8th edition, it was Stage IA (p T1 N0 M0 / ER + / HER2 Neu - / LVI & PNI – absent). Case was re-discussed in our tumor board for adjuvant treatment. She had been advised only Hormonal therapy in view of non-invasive papillary carcinoma without nodal spread. She had been started on tamoxifen 20mg. She is in periodic follow up with us as per our institutional follow up protocol and after one year of completion of her treatment, she is disease free.

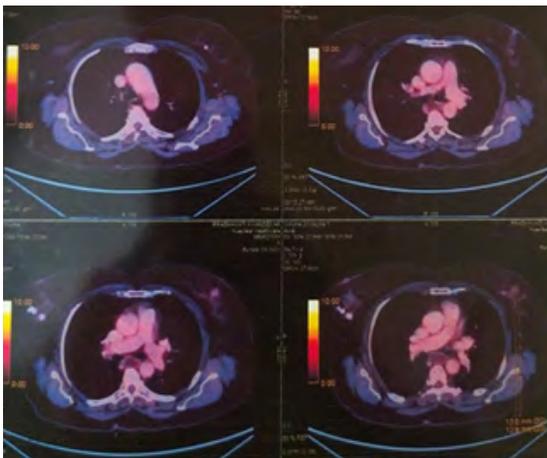


Figure 1: PET CT images of right breast lesion

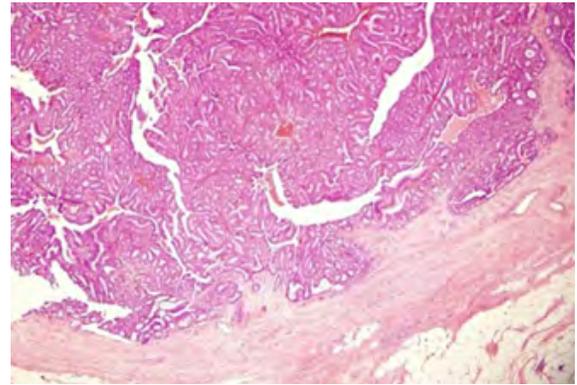


Figure 2: Microscopic examination of EPC

4. Discussion

The most common presentation of this carcinoma is with circumscribed friable mass within a cystic cavity; hence it is named as intra-cystic papillary carcinoma. It is also called as Encapsulated papillary carcinoma. It occurs most commonly in elderly population with an age range of 60-65 years [12]. It accounts for 0.5-1 % of all breast cancers [13]. Traditionally they have been classified as non-invasive tumors however, due to absence of myoepithelial cells in the tumor–stromal interface, these are considered as invasive tumors supported by recent literature [14]. In 2012 the WHO classified encapsulated papillary carcinoma as a subgroup of papillary breast lesions and a type of breast papillary carcinoma [15]. The size of the tumor ranges from 1–10 cm [16]. The classification of papillary lesions is as follows

Benign papillary lesions

1. Intraductal papilloma (solitary)
2. Intraductal papillomatosis

Atypical papillary lesions

1. Intraductal papilloma with atypical hyperplasia
2. Papilloma with DCIS

Malignant papillary lesions

Noninvasive:

1. Papillary ductal carcinoma in situ
2. Encapsulated papillary carcinoma
3. Solid papillary carcinoma

Invasive:

1. Invasive papillary carcinoma
2. Invasive micropapillary carcinoma

Their clinical presentation is with nipple discharge, a palpable mass or an asymmetric lesion [17]. Ultrasound or mammogram are not diagnostic for these types of cancer however, they can help in planning the treatment [18]. Mammography has a sensitivity and specificity of 78% and these increase to 91% when ultrasound is used along with it [19]. MRI findings have not been well documented for these cancers however, it has proven helpful for detec-

tion of DCIS around the lesion. In the index case, sono-mammography and PET-CT was done and both were non diagnostic. Histopathologically, their presentation is with absence of myoepithelial cells in the periphery of the lesion and a fibrous capsule which separates it from the rest of the stroma [10]. The index case was diagnosed only after core needle biopsy. Encapsulated Papillary Carcinoma (EPC) can be associated with DCIS or invasive ductal carcinoma [15]. In the present case, it was a non-invasive lesion and EPC have an excellent prognosis in the absence of invasive elements with a mean survival rate of 60–95% for in situ cancer [17]. The frequency of lymph node involvement is up to 11%. The frequency of distant recurrence is up to 4%, whereas frequency of local recurrence goes up to 70% [16].

The standard and documented treatment for EPC is wide local surgical excision with negative surgical margins to avoid unnecessary mastectomy. But, in the present case, consent was given for mastectomy, hence we proceeded with MRM. When EPC is associated with DCIS or an invasive component, axillary lymph node dissection is considered along with consideration of adjuvant treatment. [47]. We did level II axillary lymph node clearance. Hormonal therapy is considered useful adjuvant treatment as EPC has a high frequency of positive estrogen and progesterone receptors. The index case had been started on adjuvant hormonal therapy. The role of radiotherapy remains unclear.

5. Conclusion

Encapsulated papillary carcinoma is a rare breast carcinoma. Lack of literature support and knowledge may lead to over diagnosis and over treatment. It is difficult to differentiate between in situ and invasive cancer. It is team work of pathologist, radiologist, and oncologist in diagnosing this difficult and rare case.

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