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PAPILLARY CARCINOMA OF THE BREAST IN A PREMENOPAUSAL LACTATING WOMEN MIMICKING AS GALACTOCELE- A RARE CASE PRESENTATION AND REVIEW OF LITERATURE

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ABSTRACT

Papillary breast cancer is an extremely rare ductal malignancy, accounting for <1% of all breast cancers. Post-menopausal women are most commonly affected, while being almost non-existent in young women. It is usually an outcome of untreated in-situ ductal carcinoma. Papillary CA can be intraductal, infiltrating, or invasive. Prognosis is excellent with a 10-year survival rate in 80-90% of patients. With respect to the diagnosis, ultrasound is the most extensively used imaging modality, although an MR Mammogram has its own utility in the diagnosis. An FNA biopsy helps differentiate it from a benign isolated papilloma, which elicits similar symptoms. Various treatment modalities are being used although a segmental mastectomy is most common, followed by hormonal or radiation therapies. In the following paper, a case of a pre-menopausal lactating woman, who complained of a swelling in the left breast since 2 years, and diagnosed with papillary carcinoma.(mimicking a galactocele). A review of literature on Papillary breast carcinoma follows.

KEYWORDS: Papillary carninoma, breast cancer, papilloma, mastectomy.

INTRODUCTION

Papillary breast carcinomas represent about 0.5% of all invasive breast cancers. The name comes from the characteristic finger like small projections or 'papules' (grade 2). Similar structures arise in benign ductal papillomas and are differentiated from malignancy by the presence of myoepithelial cells lining the growth, in addition to luminal epithelial cells which are common to both. Clinically, the patient typically presents with a bloody discharge per nipple and/or mass in the breast, but if the papilloma is intracystic, there is no nipple discharge. About half of these tumors arise in the retro-areolar or sub-areolar region of the breast. Since the density of breast tissue is highest in the upper outer quadrant, most carcinomas arise there. Axillary nodes are usually not palpable.

On immunohistochemistry, these tumors are ER+/PR+ and HER2/c-erB2 negative. Since these tumors are hormone sensitive, their size is prone to variation through the menstrual cycle. Almost all cases show positive staining for Chromogranin and about 40% are also positive for Synaptophysin.

Due to lack of outcome related studies, a definite protocol on evidence based treatment options is lacking, but surgical excision remains the mainstay in majority. Hormonal therapy is added if tumor is receptor +ve.

Adjuvant therapy is being used in many centers but evidence of their role in improvement of prognosis is still lacking. Available data suggests improved outcome for papillary carcinoma as compared to invasive ductal carcinoma.

CASE REPORT

A 22 year old female patient presented with a lump in the left upper outer quadrant of the breast, which she noticed 2 years ago. The size of the swelling gradually increased during pregnancy associated with recent retraction of the nipple. It was not associated with abnormal nipple discharge. She noticed pain a few days prior to her presentation to the hospital.

Physical exam revealed an 8.0*7.5 cm lump involving the left upper outer quadrant and the central quadrant, firm, with well-defined borders, smooth surface, non-tender mass without any skin changes and no axillary lymphadenopathy.

FNA biopsy revealed a hemorrhagic pus aspirate and showed proteinaceous background with benign looking ductal epithelial cells along with sparse mixed

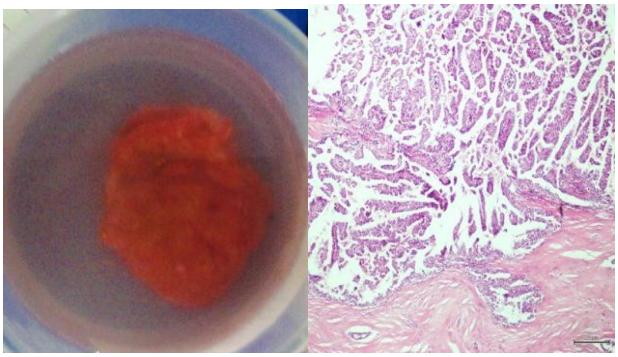
inflammatory cells consisting of neutrophils, lymphocytes and cyst macrophages suggestive of fibroadenoma with infected galactocele. A course of oral antibiotics was given.

USG Breast (Sonomammogram) of the left breast revealed a heterogenous-hypoechoic to isoechoic encapsulated lesion measuring 7.0*6.7*4.7 cm with internal vascularity in the upper outer quadrant and retro-areolar region, suggestive of benign breast disease (eg.Fibroadenoma). Rest of the parenchymal echo texture was normal without axillary lymphadenopathy.

Based on the aforementioned history and findings, surgical excision of the lump was performed and the specimen was sent for histopathological examination.
Biopsy revealed a partially solid and cystic mass of about 8.5*8.0*7.0 cm in size. Microscopy revealed Grade 1(modified Scarf Bloom Richardson's grading score 4), well differentiated invasive papillary carcinoma (pure form). Immunohistochemistry showed ER positive, PR positive and Her@/c-erbB-2 negative.

A completion modified radical mastectomy was done later and the specimen showed no residual tumor or involvement of any lymph nodes. Patient had no recurrence after 9 months of follow up. Also, tumor markers CEA and CA15-3 were within normal limits.

Patient is on regular follow up.



Gross Appearance **Microscopy**

DISCUSSION

Carter et.al in 1983 studied the clinical behavior of papillary carcinomas of the breast. They are rare malignant tumors of the breast occurring in just about 1-2% of all breast malignancies. They usually occur in older post-menopausal women, mean age being 63-67. In young women, any form of breast cancer is rare, with papillary carcinoma being almost non-existent.

Histology of this tumor shows proliferation of cells arranged around fibro vascular cones, forming a circumscribed mass. These can be divided into invasive and non-invasive varieties. Non-invasive papillary carcinomas are further subdivided into 2 types:
1. Diffuse form-papillary variant of DCIS.
2. Localized form- intracystic papillary carcinoma. (IPC, there is no nipple discharge in this variety)

It is very important to diagnose and differentiate the invasive type from the non-invasive papillary carcinoma of the breast, as each of these carry a unique prognosis.

All malignant papillary proliferations of the breast lack an intact myoepithelial cell layer within the papillae, an important feature distinguishing the malignant from benign intraductal papilloma.

On a mammogram, the presence of growth which is round, oval or lobulated, with well-defined margins raises the suspicion for malignancy. Speculations are absent.

A breast ultrasound may suggest one of the three findings:
1. An intraductal mass with or without ductal dilatation.
2. An intracystic mass
3. An intraductal mass completely filling the duct.

A galactograph/ductograph might be helpful for patients presenting with a nipple discharge.

A contrast enhanced MRI is sensitive but not specific as it is not helpful in distinguishing a benign and malignant tumor.

A mammotome biopsy is another option. It is an image guided vacuum assisted biopsy devise which can be used for clinically non-palpable lesions, but detected on mammogram. Compared to needle aspiration biopsy, it is less invasive and is associated with fewer complications, but is equally effective.

Some controversy exists regarding the optional management of these tumors. A core needle biopsy may be carried out, but for clinically and histologically worrisome tumors, surgical excision should be considered.

According to fayanju et.al, the role of both radiotherapy and hormonal therapy remain controversial. There is currently no role for cytotoxic chemotherapy.

Due to rarity of these papillary based tumors, well designed outcome studies that propose an evidence-based treatment, have not been carried out so far.

Sentinel node biopsy may be an excellent alternative to full axillary dissection in a patient with IPC and associated invasive carcinoma.

CONCLUSION

Papillary carcinoma of the beast is a rare malignancy of postmenopausal women, with an excellent prognosis. It is usually an outcome of untreated Ductal Carcinoma In Situ.

The mainstay of diagnosis includes a mammogram and a breast ultrasound, although other modalities like Galactogram, MRI, FNAC and MBS are helpful.

The treatment of tumor is based upon aspects as- features of tumor, types of cells, tumor grade (1,2 or 3), hormone receptor status, HER2 status.

Due to rarity of the tumor, well designed outcome studies on evidence based treatment have not been carried out, although surgical excision remains then mainstay of treatment in majority. The role of adjuvant therapy has not been established, although hormonal therapy may help in receptor sensitive tumors.

The overall prognosis is better than for other commoner varieties of breast malignancies like invasive ductal carcinoma.

The relevance of the case is to highlight the exceptionally rare occurrence of a papillary breast carcinoma in a pre-menopausal lactating women.

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