

Case Report

INTRACYSTIC PAPILLARY CARCINOMA: A RARE BREAST CARCINOMA

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ABSTRACT

Intracystic papillary carcinoma of the breast is a rare malignancy with excellent prognosis. We present the case of a 49 year old female patient with a huge cystic mass in her left breast which was found to be an intracystic papillary carcinoma.

Key Words: Papillary Carcinoma, Intracystic, Breast

INTRODUCTION

Intracystic papillary carcinoma is a rare malignancy of the breast accounting for 0.5 – 2 % of all breast carcinomas (Burton 2009). It has a slow growth rate and an excellent prognosis (Dogan 2003), the ten year survival is reported to be 100% (Lefkowitz, 1994). We present the case of a 49 year old woman with intracystic papillary carcinoma of the breast who presented with a huge palpable cystic mass of five years duration.

CASES

A 49 year old premenopausal woman presented with a huge lump in the left breast of five years duration. It was initially small in size which gradually enlarged to its present size. She had no nipple discharge and no family history of breast carcinoma. Physical examination revealed two large cystic lumps in the breast attached to one another. The first lump was 15 X 8 cm in size and yielded a haemorrhagic fluid on aspiration. The second lump was 6 X 4 cm in size and was found to contain a yellowish fluid on aspiration. There was no evidence of axillary lymphadenopathy. Cytopathology from the cystic fluid was negative for malignancy. Ultrasonography showed cystic masses with solid components.



Figure 1: Large cystic mass occupying nearly the whole of the breast

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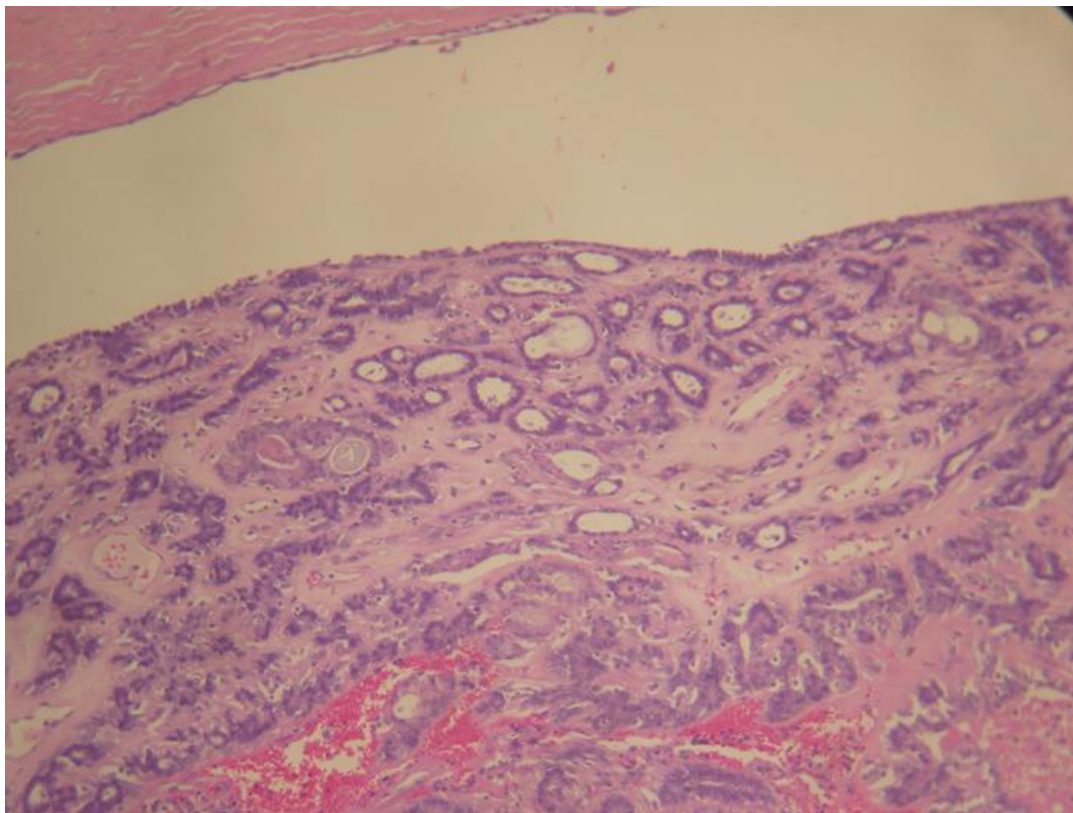


Figure 2: Left above shows collagen tissue i.e. cyst wall. Lower picture shows multiple irregularly arranged ducts of multiple sizes lined by highly irregular atypical cell, along with hyperchromatic nuclei with scanty cytoplasm, with atypical mitotic fig. & dilated congested vessels with haemorrhage and stroma infiltrated by malignant cells with calcification (primary site for psammoma body formation) -- taken on scanner view

Surgery revealed two large cystic masses attached to one another and occupying nearly the whole of the breast (Fig. 1). The whole mass was removed and sent for biopsy. Cut section showed solid outgrowth from the cyst wall. Histopathology revealed multiple irregularly arranged ducts of multiple sizes lined by highly irregular atypical cell, along with hyperchromatic nuclei with scanty cytoplasm, with atypical mitotic figures and dilated congested vessels with haemorrhage and stroma infiltrated by malignant cells with calcification (primary site for psammoma body formation) (Fig. 2). Modified radical mastectomy was done. There was no histopathological evidence of metastasis in the axillary lymph nodes. She was further treated with radiotherapy and hormone therapy.

DISCUSSION

Papillary carcinoma of the breast is distinguished by the papillary structural design: proliferation characterized by finger-like projections or fronds composed of central fibrovascular cores covered by epithelium, without myoepithelial cell layer (which differentiates between benign and malignant papillary lesion) (Benkaddour, 2012). It can be divided into invasive and noninvasive forms (Benkaddour 2012). Noninvasive papillary carcinomas are further subdivided into two subtypes: a diffuse form, the papillary variant of ductal carcinoma in situ, and a localized form, intracystic papillary carcinoma (IPC) (Benkaddour, 2012; Solorzano, 2002 and Muttarak, 2005). This localized form, describes a solitary tumor in an encysted or dilated duct (Benkaddour, 2012).

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IPC which may be unifocal or multifocal, has a propensity to occur in postmenopausal women. Women with intracystic papillary carcinoma may have no symptoms, a palpable mass, or may present with bloody nipple discharge (Dogan, 2003).

Ultrasonography usually reveals a cystic mass, with or without septations, with solid papillary masses projecting into the cyst (Knelson, 1987 and Schneider, 1989). Ultrasonography may be useful for showing wall thickening and adjacent anechoic and hyperechoic areas that may represent hemorrhage resulting from ruptured capillaries within the cyst wall or hemorrhagic infarction of the tumor cells (Dogan, 2003 and Soo, 1995). On mammography, papillary carcinoma is seen as a round or oval lobulated mass (Muttarak, 2005). If the tumor extends through the wall of the cyst, the borders may appear “shaggy” (Burton, 2009 and Estabrook, 1990).

Fine-needle aspiration (FNA) usually is highly cellular with complex papillae, foamy macrophages, and nuclear hyperchromasia and stratification. However, it is difficult to distinguish benign versus malignant papillary disease on FNA because there are no reliable and consistent features (Mugler, 2007). Also, with FNA and core needle biopsies one often obtains samples from the center of the mass but invasion is often seen at the periphery, making it difficult to differentiate between an in situ and invasive lesion (Burton, 2009; Dogan, 2003 and Mugler, 2007). Most commonly, the diagnosis of IPC occurs after excisional biopsy. Gross appearance of IPC may show a cyst surrounded by a fibrous wall containing dark brown blood clots (Burton, 2009 and Czernobilsky, 1967).

Fortunately, IPC is a slow growing tumor with a good prognosis. Historically, IPC was treated with radical mastectomy. However, recurrence rates following treatment with only wide local excision appear to be low (Burton, 2009; Mugler, 2007 and Czernobilsky, 1967). Wide local excision of the breast lesion with or without sentinel node biopsy can be done for pure intracystic papillary carcinoma. Axillary dissection is not required since nodal metastasis is rare in this group of patients, however since there have been a few reports of nodal metastases, sentinel node biopsy may be warranted (Deshmukh, 2012 and Mulligan, 2007). Mastectomy is done for large tumors (where breast conservation is not possible), for tumors with associated intraductal component and those with frank invasion (Burton, 2009 and Deshmukh, 2012). Further management for these groups of patients is radiation therapy and hormone therapy. Hormone therapy is required in a majority of these patients as they are mostly ER and PR positive (Deshmukh, 2012).

To conclude, intracystic papillary carcinoma is a rare breast malignancy with excellent prognosis. The 10-year survival rate has been reported to be 100% (Lefkowitz, 1994). Awareness of this rare entity helps in better management of these patients.

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