Intracystic papillary carcinoma of the breast in a 21-year old premenopausal Nigerian woman: a case report

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Abstract

We report the case of a 21-year-old Nigerian woman who presented to us with features of intracystic papillary carcinoma, a rare form of breast cancer usually seen in postmenopausal women in their sixth to eighth decades of life. To the best of our knowledge, there has been only one other case report of this lesion occurring in women in their second decade of life.

Physical examination showed a well-defined mass, 54 mm in diameter, in the upper proximal quadrant of the right breast close to the areola, histologically composed of monotypic epithelial cells disposed in solid, cystic, and papillary patterns. A diagnosis of intracystic papillary carcinoma was made because of the presence of intracystic arteriovenous malformation of the fibrovascular stroma, a monotonous cell population, the presence of mitoses, and the lack of myoepithelial cells determined by immunohistochemistry using calponin and p63 stains. Estrogen receptor status was positive while progesterone status and HER-2/neu receptor status were negative.

The patient has survived for 12 months without any sign of recurrence after the last surgical resection of the tumor.

Introduction

Intracystic papillary carcinoma (IPC) is a variant of intraductal papillary carcinoma, a rare histological type of breast cancer occurring only in about 1-2% of women.1,2 Papillary carcinomas can be divided into invasive and noninvasive forms. Intracystic (encysted) papillary carcinoma is the localized, noninvasive form, occurring as a grossly evident tumor in a cystic and dilated duct.3 The patterns seen include micropapillary, cribriform, trabecular, and solid. Occasionally they may be mistakenly diagnosed as papillomas.4

Because of its rarity, there is a paucity of literature on IPC in women younger than thirty years old, as it predominantly affects elderly, postmenopausal women.5 It has been documented in males as well.6 To the best of our knowledge, there has been only one other case report of this lesion occurring in premenopausal women in their second decade of life.7 We recently encountered a case of low nuclear grade intracystic papillary carcinoma of the right breast in a 21-year-old woman, which is probably the first report in our environment.

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Clinical history

A 21-year-old woman presented in our surgical out-patient department with a four-year history of a right breast mass and bloody nipple discharge. Her family history was negative for breast cancer and there was no history of exposure to antinecancer drugs, radiation, or cigarette smoking. Clinical examination confirmed a well-defined 54-mm mass in the upper proximal quadrant of the right breast close to the areola, with an ipsilateral bloody nipple discharge (Figure 1). There was no associated axillary lymphadenopathy.

Mammography was not performed as facilities for this are unavailable in the state. Core needle biopsy was performed using a 23-gauge needle and showed a papillary tumor histologically composed of monotypic epithelial cells. The preliminary diagnosis was atypical ductal papilloma with a differential diagnosis of intracystic papillary carcinoma. Four weeks after the core biopsy, excision biopsy of the tumor was performed.

Pathological features

Grossly the excised tissue showed a mass of fibro-fatty tissue weighing 101 g and measuring 80x60x12 mm. The cut surface of the tissue showed a well-defined mass approximately 17 mm in diameter, with a cystic cavity filled with pale, friable, papillary tissue. Histologically the tumor was composed of closely packed epithelial cells (nuclear Grade 2) with a papillary architecture within a cyst (Figure 2). Biopsy site changes were apparent in the surrounding tissue.

Immunohistochemical (IHC) stains for calponin, p63, and CK5/6 were performed on the tumor and demonstrated the absence of a myoepithelial cell border in the papillary cores as well as at the periphery of the lesion. Based on these findings, the diagnosis of intracystic papillary carcinoma was made. Hormone receptor analysis showed mild estrogen receptor positivity (1+) while both progesterone and HER-2/neu were negative.

The patient had another excision 15 weeks later because of involved margins, which revealed residual papillary lesions as well as a few foci of mucinous carcinoma. Based on this associated pathology and her age, we considered adjuvant radiotherapy appropriate for her but opted for tamoxifen when her family expressed their inability to cope with the inconvenience and expense of radiotherapy in another center. Although she is yet to comply with hormonal therapy for poorly understood reasons, she has been symptom-free for about a year.

Discussion

Typically patients with IPC present with a palpable mass (80% or more) and/or bloody nipple discharge (up to 22%).8 Less frequently they may present as a radiographic abnormality.9 Most patients are postmenopausal women usually with median ages ranging from 63-75 years.10 Its occurrence in this 21-year-old was a novelty. The duration of symptoms in patients with IPC varies but prolonged periods of one year or more is not considered unusual.11 Late presentation, as seen in our patient who had a four-year history, is a typical feature of most patients with breast cancer in sub-Saharan Africa, in contrast to what occurs in

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screening mammography because of their patients like ours usually are not subjected to accurate diagnosis in patients with multicentric tumors. Excision biopsy is recommended for pathologic diagnosis because of the frequently inconclusive findings associated with fine needle aspiration or core needle biopsy alone. Immunohistochemistry with stains such as calponin, smooth-muscle myosin heavy chain (SMM-HC) cytoplasmic stains, and p63 nuclear stains have been invaluable in distinguishing IPC, which does not appear to have a myoepithelial cell layer around tumor nodules, from ductal carcinoma in situ (DCIS). Another differential diagnosis, papilloma, can be distinguished from IPC using the criteria proposed by Kraus and Neubecker. Usually IPC presents with low to intermediate grade, high estrogen and progesterone receptor positivity, and negativity for c-erb-2.

There are no well-defined views on the management of IPC; however, surgical excision is the mainstay of treatment. In selected studies, patients with IPC were divided into three groups (IPC alone, IPC with associated ductal carcinoma in situ, and IPC with associated invasion with or without ductal carcinoma in situ), and the prognosis did not differ among the three groups regardless of the type of surgery performed (breast-conserving surgery or mastectomy) and whether adjuvant therapy was administered. Fayanju et al. observed that patients with IPC and associated DCIS or microinvasion were treated with adjuvant radiation and endocrine therapy on the basis of this associated pathology. Therefore sentinel lymph node biopsy has been advocated for these patients owing to the low rate of axillary lymph node metastases (8-11%) observed in those evaluated for regional disease.

Sentinel lymph node biopsy also seems justified in large IPC tumors owing to the occasional axillary lymph node involvement observed in a few cases of pure IPC characterized by lesions measuring 4 cm or more. Pure IPC has been reported to have an excellent prognosis even when treated with local excision only, but the current clinical management remains variable with a potential for overtreatment.

Only in a subset of younger patients (<50 years old) with pure IPC was adjuvant radiotherapy and/or hormonal therapy administered and this is recommended.

The prognosis of IPC, particularly in elderly women, is excellent with few or no cancer-related deaths in most study series. In contrast to the lower relative survival rate for patients with non-IPC cancers such as invasive ductal carcinoma, even patients with invasive IPC had a survival rate of >90% of that of the general population. Factors such as age at presentation, tumor diameter, and nuclear grade, necrosis, and the presence of associated lesions such as invasion and DCIS, all affect patient survival.

In summary, intracystic papillary carcinoma is extremely rare in women in their second decade of life but it does occur. The diagnosis depends on finding typical clinical features, radiologic and pathologic investigations and, invariably, immunohistochemistry. The tumor should be managed with or without adjuvant therapy on the basis of any associated pathology, tumor size, and age of the patient. Although our patient has been symptom-free for one year now, the prognosis in this age group is yet to be evaluated.

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