

Intracystic Papillary Carcinoma of the Breast

Élan C. Burton, M.D., Kurt R. Stahlfeld, M.D., FACS, and John A. McKeating, M.D., FACS

We present the case of a 63-year-old woman with intracystic papillary carcinoma of the breast who presented with a palpable mass, one year after a negative mammogram. Sonography showed a complex mass and ultrasound-guided aspiration cytology was negative, and the patient returned 6 months later with a recurrent mass. Excisional biopsy revealed invasive intracystic papillary carcinoma. Intracystic papillary carcinoma is a rare malignancy of the breast primarily affecting postmenopausal women. As opposed to invasive micropapillary carcinoma, intracystic papillary carcinoma is a low-grade carcinoma with a favorable prognosis.

Introduction

Intracystic papillary carcinoma is a rare malignancy of the breast primarily affecting postmenopausal women. Classification and treatment strategies frequently subdivide patients with intracystic papillary carcinoma into three equally prevalent subsets: those with intracystic papillary carcinoma alone, those with intracystic papillary carcinoma and accompanying ductal carcinoma in-situ, and those with evidence of invasion. As opposed to invasive micropapillary carcinoma, intracystic papillary carcinoma is a low-grade carcinoma with a favorable prognosis. Treatment modalities including the extent

of surgical excision, lymph node dissection, radiation, and chemotherapy are determined by the grade and size of the lesion and the associated pathology. We present the management dilemmas in a patient with intracystic papillary carcinoma, ductal carcinoma in-situ, and three positive axillary lymph nodes.

Case Report

A 63-year-old woman presented with a recurrent mass six months following aspiration of a complex mass in her right breast. Annual screening mammography initially revealed no abnormalities (Figure 1). However, one year later a diagnostic mammogram was obtained secondary to the patient developing a palpable mass. A well circumscribed complex mass was found at the 5 o'clock position that correlated with the palpable mass classified as a Bi-RADS 4. At that time, an ultrasound guided aspiration of the cyst yielded fifteen milliliters of cloudy, partially bloody fluid. As cytopathology was negative for malignant cells, the patient was advised to follow-up in six months with no intermediate intervention. At her six month follow-up, patient noted that

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Abbreviations: CC, craniocaudal; IPC, intracystic papillary carcinoma; MLO, mediolateral oblique

Élan C. Burton, M.D. (Email: ecbduke2000@hotmail.com), Kurt R. Stahlfeld, M.D., FACS, and John A. McKeating, M.D., FACS, are in the Department of Surgery, Mercy Hospital of Pittsburgh, Pittsburgh, PA, United States

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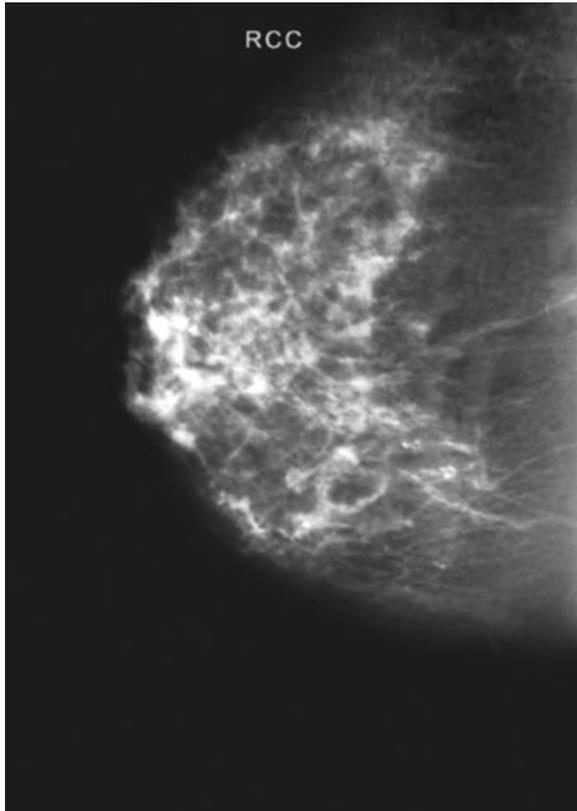


Figure 1. 63-year-old woman with intracystic papillary carcinoma of the right breast. Right CC view showing no abnormality one year before presentation.

the mass had returned and mammogram again showed a complex mass classified as a BI-RADS 4. Since the patient previously underwent aspiration with return of the mass, the patient elected to have excision of the mass for a more definitive treatment. Her past medical history was notable for breast cancer in two first degree relatives and a previous right sided breast biopsy for focal ductal ectasia and bloody nipple discharge. On physical exam, a freely mobile 2.5 cm mass was palpated in the right breast at the site of the previous aspiration. No axillary or supraclavicular nodes were palpable. Mammography (Figure 2 and 3) and ultrasound (Figure 4) confirmed a 2.5 cm partially circumscribed complex cyst with a solitary punctate calcification. Excisional biopsy was performed and pathology noted a 3 cm cyst with invasive intracystic papillary carcinoma and accompanying cribriform ductal carcinoma in-situ (Figure 5).

Immunoperoxidase stains were strongly positive for estrogen receptors, moderately positive for progesterone receptors, and negative for HER 2 NEU. Sentinel lymph node biopsy revealed three lymph nodes positive for adenocarcinoma. The remainder of her work up was negative for metastatic disease. She was treated with radiation therapy, 6 cycles of docetaxel, doxorubicin, cyclophosphamide, and five years of anastrozole. After extensive discussion but without genetic testing, the patient elected to undergo bilateral prophylactic mastectomies, total abdominal hysterectomy, and bilateral salpingo-oophorectomy. She remains disease free 20 months following her resection.

Discussion

Intracystic Papillary Carcinoma (IPC) is a rare malignancy of the breast accounting for 0.5% - 2% of all breast carcinomas. It occurs more frequently in postmenopausal women over the age of 60. At presentation, the patient may note a painless, mobile mass and complain of bloody nipple discharge. Mammography typically shows a large circumscribed mass with an irregular or nodular contour [1-3]. If the tumor extends through the wall of the cyst, the borders may appear “shaggy” [2]. On ultrasound, there is usually a cyst with papillary fronds in the cyst cavity [2,4-5]. Septations may or may not be present [2,4-5]. Fine-needle aspiration (FNA) usually is highly cellular with complex papillae, foamy macrophages, and nuclear hyperchromasia and stratification [1]. However, it is difficult to distinguish benign versus malignant papillary disease on FNA because there are no reliable and consistent features [1]. 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 [1,4]. Most commonly, the diagnosis of IPC occurs after excisional biopsy [1,4]. Gross appearance of IPC may show a cyst surrounded by a fibrous wall containing dark brown blood clots [6]. Histologic features include arborization of fibrovascular stroma covered by a single or multiple layers of cells and a lack of myoepithelial cells [4,7-8].

Fortunately, IPC is a slow growing tumor with a good prognosis. Historically, IPC was treated with radical mastectomy [1,6]. However, recurrence rates following treatment with only wide local excision ap-

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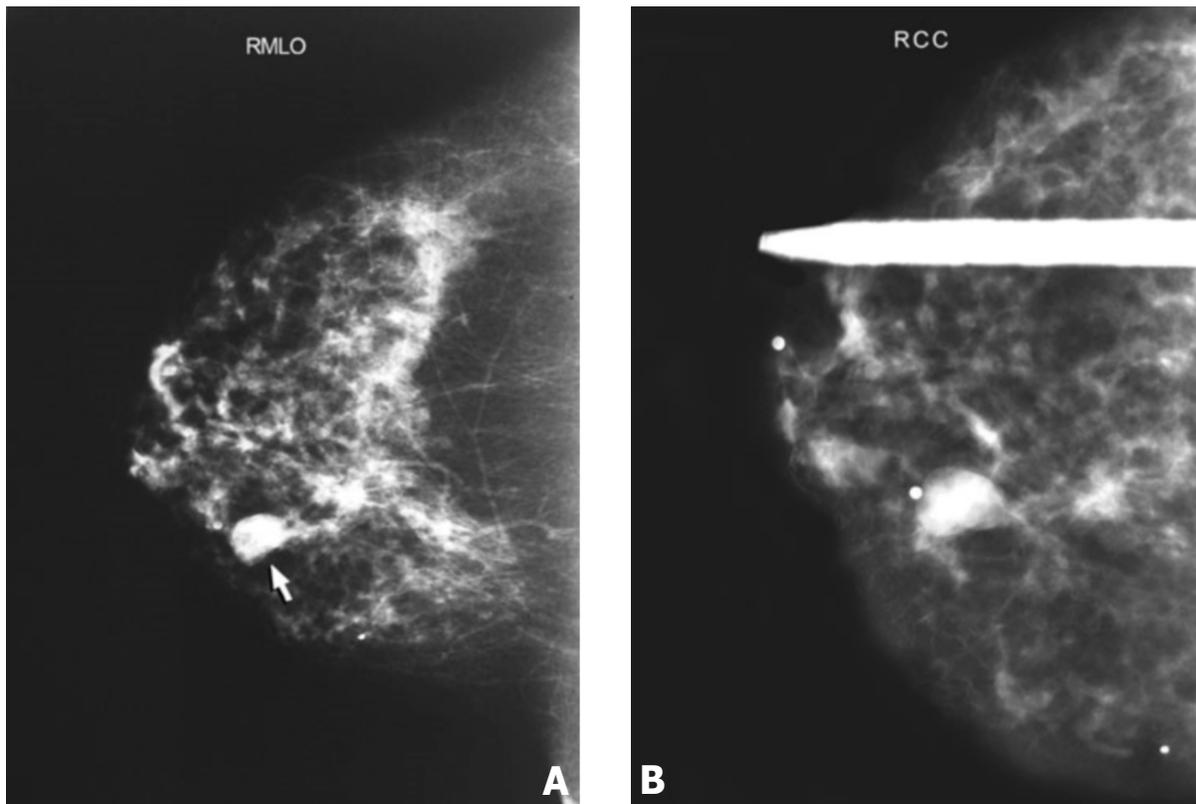


Figure 2. 63-year-old woman with intracystic papillary carcinoma of the right breast. **A**, Right CC view of an anterior well circumscribed bilobed dense mass (arrow) at the five o'clock position. **B**, Right CC view spot magnification showing a BB overlying the mass (arrow).

pear to be low [1,9]. Since less than one percent of IPC without ductal carcinoma in-situ (DCIS) is associated with lymph node metastasis, Harris recommends treating pure IPC with wide local excision without axillary lymph node dissection [1,9]. However, IPC may also be associated with DCIS or have areas of invasion. Given that the presence of DCIS in patients with IPC correlates with a higher incidence of local invasion and lymph node involvement, these patients and those with definite invasion should be treated with sentinel lymph node dissection [1,8]. The extent of the local resection margin and the role of mastectomy is not yet defined [1,4]. The need for radiotherapy and chemotherapy in IPC, IPC with DCIS, and IPC with invasion also remains unclear [1,8]. Reported rates of recurrence among these three subsets vary significantly, independent of the extent of surgery and use of adjuvant radiation [1,8]. Fortunately,

many IPC tumors are estrogen and progesterone receptor positive, suggesting a more favorable diagnosis and benefit from hormone guided therapy [1,9].

Our patient was ER and PR positive, had IPC with concurrent DCIS, and had metastatic deposits in three axillary lymph nodes. Following breast conserving therapy, radiation, and chemotherapy, she elected to have prophylactic mastectomies. Although she remains disease free, her course and treatment demonstrate that specific treatment guidelines for IPC in combination with DCIS or invasion must be individualized.

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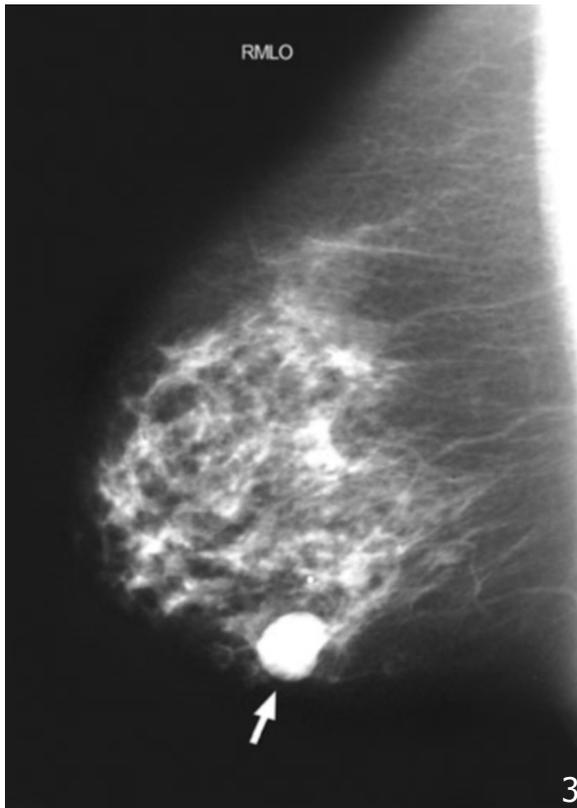
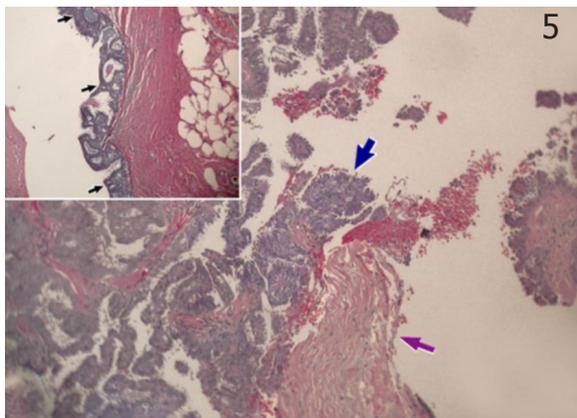


Figure 3. 63-year-old woman with intracystic papillary carcinoma of the right breast. Right MLO view of a well circumscribed mass (arrow).



Figure 4. 63-year-old woman with intracystic papillary carcinoma of the right breast. Sonogram showing a thick walled irregular complex mass with several septations and an echogenic component (arrow).



< Figure 5. 63-year-old woman with intracystic papillary carcinoma of the right breast. Hematoxylin and eosin stain demonstrating malignant cells (blue arrow) penetrating through the cyst capsule (purple arrow) into the surrounding fat. Inset shows an area of concomitant DCIS (small arrows).

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