# Case Report

# Intracystic Papillary Carcinoma of the Breast: Risks in Diagnostic, Staging and Treatment

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#### Abstract

Intracystic papillary carcinoma (IPC) of the breast is a rare form of breast in situ carcinoma that comprises 0.5-2% of all breast tumors. IPC of the breast occurs in women around the sixth decade of life and presents an injury of great dimensions, more frequently retroareolar. Clinical and radiological findings can be confused with advanced-staged invading carcinomas (T2 or T3), leading to unnecessarily aggressive treatment. The correlation of clinical, radiological and anatomopathological findings is necessary for a correct diagnosis. We present two cases of IPC and discuss diagnostic characteristics.

Keywords: Breast Cancer Papiloma. Papillary Carcinoma. Invading Carcinoma. Staging.

## Introduction

Papillary injuries of the breast include injuries of peripheral ducts (micropapilomas) and injuries of subsegmentary or galactofore ducts (intraductal papilomas). In these injuries, all the specter of proliferation of the known ductal epithelium in the breast can occur, such as hyperplasia without atypias, with or without apocrine metaplasias, atypical hyperplasias and in situ carcinomas. Papiloma-originated in situ carcinomas also are known as intracystic papillary carcinomas, for they occur in large caliber mammary ducts, enlarged by the presence of the injury. Eventually, foci of invasive ductal carcinoma develop from these in situ injuries, invading stroma adjacent to compromised ducts. Intracystic Papillary carcinoma of the breast (CPI) is a rare form of not invasive tumor of the breast and represents 0.5%-1% of all breast neoplasias.<sup>1-5</sup> It is generally a long evolution

tumor located in subareolar region<sup>1,3,5</sup> and normally affects aged women (average 65 years). Clinically, it can be identified by the presence of a palpable tumor, papillary stroke, and skin retraction.<sup>1</sup> The size can vary from some millimeters to some centimeters causing difficulties diagnostic and clinical staging. As regards diagnosis, some resources can be used such as ultrasound, mammography and fine needle aspiration biopsy (FNAB).<sup>1</sup> Surgical and adjuvant treatments are used considering staging and the different anatomoclinical factors, but prognostic is generally very good.<sup>1,3,4,5</sup>

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#### **Case Report**

#### Case 1

A patient of 82 years of age presenting a hardened, mobile palpable injury with defined contours, measuring 6 x 5cm, in the superior-lateral quadrant of the right breast, without axillary lymph node megaly. Mammography showed a dense, heterogeneous 4.0cm image (BI-RADS IVc) and ultrasound described a solid nodule with 3.5 cm. Fine needle aspiration biopsy (FNAB) suggested a papillary tumor. FNAB diagnosed in situ ductal carcinoma with a focus of invasion. The patient was submitted to total right mastectomy with axillary emptying (Level I), in January 2006. Anatomopathological examination of the surgical piece showed intracystic papillary carcinoma without foci of invasion or compromising in the 13 dissected lymph nodes (Figures 1, 2 and 3). Complementary adjuvant treatment was unnecessary.

#### Case 2

A patient of 65 years of age presenting a palpable retroareolar nodule of 5.5cm that to mammography presented a dense, lobulated nodule of badly defined contours, measuring 4cm (Figure 4). With ultrasound a solid-cystic nodule with intense peripheral vascularization was observed (Figure 5).



**Figure 1** – Macroscopic aspect of intra-cystic papillary carcinoma of the breast. The injury is well delimited and presents an enlarged mammary duct having a papillary injury on which in situ carcinoma originated



**Figure 2** – Panoramic microscopic aspect of the intracystic papillary carcinoma of the breast. Papillary injury is inside mammary duct with a thickened wall due to fibrosis. The anatomopathological study of this nodule did not identify tumor points of invasion beyond the duct wall



**Figure 3** – Detail of the architecture of intracystic papillary carcinoma. The ramified conjunctive-vascular axis (white star) is coated by cell proliferation with a cribriform architecture and nuclear monotony

FNAB and *core-biopsy* identified a papillary injury with atypias and without an invasive component. Due to this result a conservative surgical conduct (quadrantectomy) was chosen. The anatomopathological study showed an intracystic papillary carcinoma measuring 4 cm, with a 1.5 cm focus of invasion. Sentinel lymph node study was negative for micrometastasis, and the patient had no axillary emptying.



**Figure 4** – Case 2 – Mammographic aspect of intracystic papillary carcinoma of the breast. In this 4.0 cm nodule, anatomopathological study of the surgical piece showed an invasive focus of 1.5 cm



**Figure 5** – Case 2: Ultrasonographic aspect of intracystic papillary carcinoma. A retroareolar solid-cystic injury with intense located peripheral vascularization, presenting the conjunctive-vascular axis of the injury

#### Discussion

Intracystic papillary carcinoma of the breast is an uncommon injury, but is able to induce diagnostic and therapeutic errors if not dully identified by postoperative exams. It generally presents as an only injury of large dimensions in women in the sixth decade of life and that initially suggests locally advanced tumor injury. The reported cases demonstrate the difficulties found in dealing well with these patients.

The first patient presented inconsistence regarding FNAB and *core-biopsy*. The patient was submitted to ablative treatment due to the risk of having a locally advanced invasive ductal carcinoma. The final staging of this patient was TisN0M0 (Stage 0) for a previous clinical staging was T3N0M0 (Stage IIB).

The second was patient correctly diagnosed as having an atypical papillary injury, which resulted in a conservative treatment. The study of the surgical part identified an invading component of 1.5cm. In this case, final staging was T1N0M0 (Stage I) for a previous clinical staging T3N0M0 (Stage IIB). One must remember that patient having invading carcinomas larger than 5cm are candidates to neoadjuvant treatment, and an inexact diagnosis, in case of the IPC, can lead to this additional unnecessary treatment.

To mammography, IPC is generally seen as an oval mass of well-defined contours, frequently located in the retroareolar region.<sup>2</sup> Ultrasound generally finds a cystic mass with or without septations with a solid content projecting inside, or simply a predominantly solid mass, and in these important cases the important factors are the characteristics of the injury wall and areas of anechogenicity or hyperechogenicity that may present hemorrhages.<sup>2</sup>

Histologically, the challenge of papillary injuries study is the identification of atypias and the meeting of the qualitative and quantitative criteria for study in situ carcinoma diagnosis, and sometimes a immunohistochemical test is needed for a better characterization of the injury's cell populations. Thick needle biopsy not always is sufficient in the initial differentiation of these injuries.<sup>5,6</sup> Final diagnosis can only be evaluated after a complete resection, since the presence of invasive tumors foci may be microscopic.<sup>2</sup> Central areas of the papillary injury can suffer ischemic necrosis and fibrosis which simulate an invasive component, but a true invasive component only occurs when there is invasion of the enlarged duct wall and of adjacent stroma. Three histopathological forms of papillary injury are described: pure intraductal papiloma, papiloma associated to in situ ductal carcinoma ISDC (IPC) and IPC associated with ISDC.5 The choice of treatment for IPC depends on the presence or absence of ISDC. Surgical procedures vary from segmentary resection with or without lymphadenectomy to total mastectomy with or without axillary emptying.1 Total mastectomy with axillary emptying allows for a long disease-free period, but it is not necessary for all patients. Carter et al.<sup>2</sup> demonstrated

that pure IPC is less aggressive and mastectomy is in most cases an extreme procedure. Carmen et al.<sup>5</sup> studied retrospectively forty patients of the three groups and observed that recurrence rate and survival did not modify according to the used surgery or the use of postoperative radiotherapy, but also observed that in IPC cases axillary emptying is not justified, because axillary lymph node compromising is rare, and indicate as an alternative for tumors associated to ISDC sentinel lymph node biopsy or the selective emptying (level I). Adjuvant chemotherapy is only indicated in the presence of axillary metastases<sup>1</sup> and hormonetherapy can be suggested depending on tumor characteristics.

IPC has therefore a good prognostic up to a 100% 10-year disease-free survival rate mainly when in its pure form, and clinical superstaging must be prevented not to induce an unnecessarily aggressive treatment.

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