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Review Article

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INTRACYSTIC PAPILLARY CARCINOMA OF THE BREAST: A RARE ENTITY (ABOUT AN OPERATED CASE AND LITERATURE REVIEW)

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ABSTRACT

Intracystic papillary carcinoma of the breast (ICPC) is a very rare entity and represents 0.5 to 1% of all breast carcinomas. It may present a misleading cystic appearance and is generally characterized by slow growth with a good prognosis. Echo-mammography guides the diagnosis, Histology often reveals a papillary carcinoma. We report a case of ICPC revealed by histologically proven axillary lymph node metastasis, in a patient treated by conservative surgery, followed by radiotherapy and hormonal therapy based on Tamoxifen offering good locoregional and distant control. The aim of this study is to recall the diagnostic, therapeutic and evolutionary aspects of this rare tumor.

KEYWORDS: Carcinoma, papillary, Intracystic, Breast.

INTRODUCTION

Intracystic papillary carcinoma (ICPC) is a very rare variant of intraductal carcinoma, constituting only 0.5% to 1% of all breast cancers. Its anatomopathological appearance is characteristic, it must be differentiated from in situ ductal carcinoma (ISDC) or invasive carcinoma, which are sometimes associated with it. ICPC is generally characterized by slow growth with a good prognosis. On the occasion of this observation, we recall the diagnostic, therapeutic and evolutionary aspects of this rare tumor.

PATIENT AND OBSERVATION

Mrs. BK, 67 years old, G14P14, postmenopausal for 30 years, poorly monitored for high blood pressure having presented a month before the consultation a right axillary swelling without any other associated sign, of which the infectious origin was first suspected (Figure 1). The patient underwent an axillary ultrasound showing an appearance of right axillary lymphadenopathy. The thoraco-abdominopelvic scanner revealed voluminous images suggestive of right axillary lymphadenopathy. The search for the expert gene to eliminate any tuberculosis origin came back negative. Axillary surgical exploration for diagnostic purposes was carried out, revealing a fixed lymph node encircling the axillary pedicle, the biopsy performed came back in favor of lymph node metastasis from a probable breast carcinoma (Figure 2). An echo-mammogram was requested revealing a nodule located in the supero-external quadrant of the right breast measuring 2 cm × 1.5 cm in diameter, with axillary lymphadenopathy (Figure 3).

After a large lumpectomy, the pathological study showed a completely excised intracystic papillary carcinoma with clear healthy margins, classified grade II according to the modified SBR classification with vascular emboli without ISDC or lobular neoplasia (Figure 4), hormone receptors were positive and Ki67 was 10%. The patient received adjuvant chemotherapy followed by axillary lymph node dissection.

The postoperative course was simple. The patient was referred to the oncology department for axillar radiotherapy and hormonal therapy. Locoregional and remote control was good.

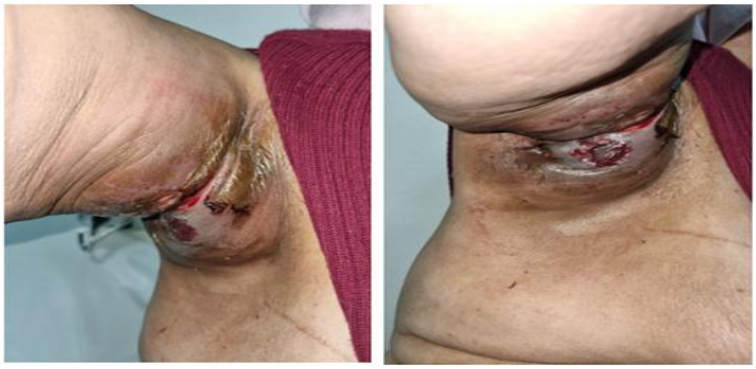

Figure 1: Right axillary lymph node mass.

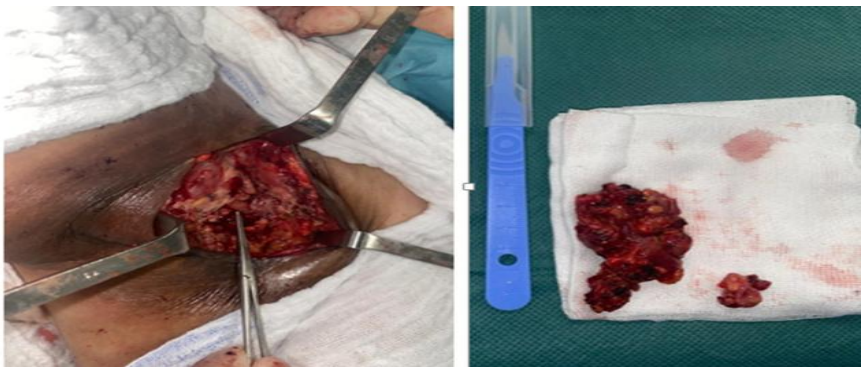

Figure 2: Lymph node biopsy image.

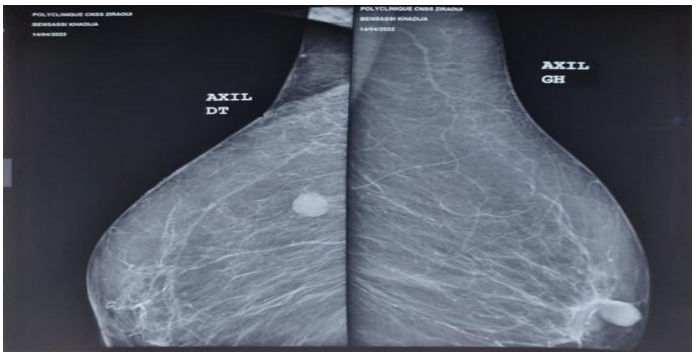

Figure 3: Mammogram showing a mass in the supero-external quadrant of the right breast.

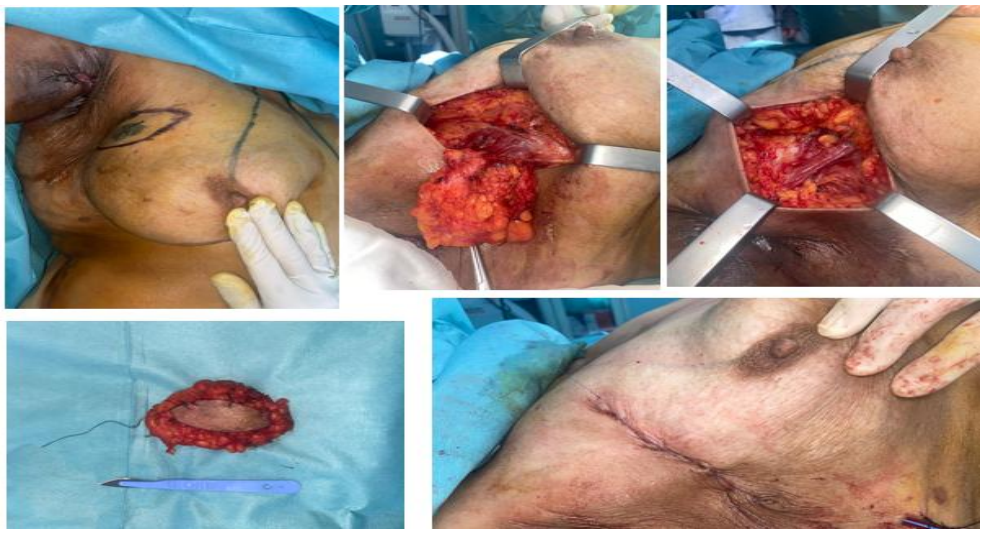

Figure 4: Intraoperative image of lumpectomy.

DISCUSSION

ICPC is a rare malignant ductal tumor, representing 0.5 to 1% of all breast carcinomas. It is a papillary lesion located in a dilated or cystic milk duct. It generally occurs after the age of 40 with an average age varying from 55 to 67 years old depending on the authors. In approximately 50% of cases it is centrally located and more precisely in the retro-areolar region. Tumor size varies from 1 to 14 cm.[1] The classic radiological sign on mammography is a sharply defined, well-circumscribed, oval or polylobed opacity. On ultrasound, it appears as a complex cystic mass with a solid component showing vascular flow on color Doppler. These radiological characteristics should raise suspicion of this rare form of breast cancer. Contrast-enhanced magnetic resonance imaging (MRI) of the breast can guide the diagnosis by showing septation and mural nodules.[2] Biopsy of the lesion involving the solid portion is generally more informative. The macroscopic study finds within a cyst, with a thick and fibrous wall, a polylobed, friable and hemorrhagic formation. On microscopy, the tumor architecture is most often papillary with cribriform appearances. The diagnosis of stromal invasion remains difficult.[3]

The therapeutic strategy is not clearly established, given the rarity of this form of breast cancer. Breast-conserving surgery with wide excision is the most used method. Carter et al.[4] from a series of 7 cases of isolated ICPC, having been treated by lumpectomy, did not observe local recurrence after a follow-up of 7 years. The absence of axillary lymph node metastasis in the study by Baron et al.[5] and that of Harris et al.[6] combined with the absence of recurrence, suggest that the treatment of choice for isolated ICPC is an enlarged lumpectomy. However, in certain cases, mastectomy with or without immediate breast reconstruction may be offered (in cases of large tumors, insufficient margins, recurrence and on patient choice). Lymph node metastases are exceptional. Axillary lymph node surgery in the form of sentinel lymph node biopsy or axillary dissection should be avoided to spare patients the morbidity of axillary dissection.[7] In our case the tumor was revealed by lymph node metastasis which justified lymph node dissection. The search for arguments to support the theory that adjuvant radiotherapy significantly reduces the risk of local relapse in patients who have had breast-conserving surgery in cases of ICPC is still ongoing. However, numerous articles and published data recommend radiotherapy in young women under 50 years of age, in forms associated with invasion and/or in situ ductal carcinoma (ISDC).[8] The low potential for metastasis and vascular invasion makes chemotherapy not obligatory. In our case the pathological tissue study returned with an invasive intracystic papillary carcinoma ICPC with vascular emboli. Adjuvant hormonal therapy mainly with tamoxifen should be prescribed to reduce the risk of local recurrence in cases of positive hormone receptors. Despite these general principles, the optimal treatment of ICPC remains controversial.[9] ICPC is generally characterized by slow growth with a very good prognosis compared to other intraductal carcinomas. Lefkowitz reports a 10-year disease-free survival rate of 91%.[10]

CONCLUSION

Intracystic papillary carcinoma is a particular and rare entity of breast cancer. Its prognosis is excellent in its isolated form. The diagnosis is suggested by ultrasound and then confirmed by histopathological and immunohistochemical examination. Therapeutic management remains variable, however treatment remains conservative in the absence of any infiltrative component. The basis of treatment remains mastectomy-type surgery or breast-conserving lumpectomy. Adjuvant radiotherapy and/or hormonal therapy should be considered in appropriate cases.

Conflicts of interest

The authors declare no conflict of interest.

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