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Rare Presentations of Encapsulated Papillary Carcinoma: A Case Report in Male Breast and a Case of Upfront Metastatic Presentation

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Abstract **Case Series**

Encapsulated papillary carcinoma (EPC) of the breast, while rare, presents unique diagnostic and management challenges. This report sheds light on two exceptional cases of EPC, diverging from typical clinical presentations. The first case involves a male patient, while the second exhibits an unusual upfront manifestation with metastasis. Through detailed clinical examinations, imaging modalities, and histopathological analyses, both cases underscore the importance of thorough evaluation and tailored treatment strategies in managing this rare malignancy. Additionally, insights from epidemiological data and treatment paradigms further elucidate the complexities surrounding EPC diagnosis and management, emphasizing the need for individualized approaches to optimize patient outcomes.

Keywords: Encapsulated Papillary Carcinoma- Male Breast cancer-Radiotherapy.

INTRODUCTION

Encapsulated papillary carcinoma (EPC) is a rare breast cancer, constituting about 1–2% of all breast carcinomas in women, and it is associated with an excellent prognosis [1]. Typically, this cancer is diagnosed in postmenopausal women [2]. However, instances of EPC have been documented in males as well. In this report, we highlight two rare clinical presentations of encapsulated papillary carcinoma. The first case involves a male patient, and the second case is characterized by an unusual upfront presentation with metastasis.

CASE 1

A 63-year-old man with a history of type 2 diabetes and two paternal cousins diagnosed with breast cancer consulted for a right breast nodule discovered through self-palpation, evolving over two years. Clinical examination revealed the presence of a 4 cm firm, mobile nodule in the infero-external quadrant, without inflammatory signs or nipple discharge. Axillary lymph nodes were free. Bilateral mammography revealed a well-defined, non-calcified oval hydric opacity in the lateral external mammary region without parietal anomalies such as thickening or flattening, and no associated nipple retraction. Ultrasound complement showed a subcutaneous, well-defined oval lesion with a tissular echostructure, measuring 32 x 18 mm, displaying areas of necrosis and a postero-inferior vascular pedicle, without regional lymphadenopathy (Image 1). A wide local excision was performed. Histological examination revealed a tumor proliferation consisting of numerous fine fibro-conjunctival axes lined by 2 or 3 layers of epithelial cells with moderate to severe atypia. These cells had rounded, slightly anisocaryotic, nucleolated nuclei, with rare mitotic figures. Focal cribriform architecture was observed. The lesion was surrounded by a thickened fibrous capsule, respected without peritumoral invasion signs. The lateral margins were healthy, with the closest at 3mm (superficial). Immunohistochemistry indicated positive, intense hormonal receptors in 100% of cells, Ki67 estimated at 25%, and P63 confirming the absence of myoepithelial cells. HER2 was negative, suggesting a high-grade encapsulated papillary breast carcinoma without capsular invasion. The thoraco-abdomino-pelvic CT scan confirmed the presence of the breast nodule without distant secondary lesions (Image 2). Further surgery with mastectomy and axillary clearance was performed, histological study showed no infiltrating carcinoma residual tumor, and no lymph node metastasis (0 N+/20N). Lateral and deep margins were healthy. A PET CT performed for the staging was normal. The patient received radiotherapy on the left thoracic wall at a dose of 40 Gy in 15 fractions of 2.67 Gy, followed by hormone

therapy with tamoxifen 20mg. The patient is in good locoregional and distant control with a follow-up of 26 months.

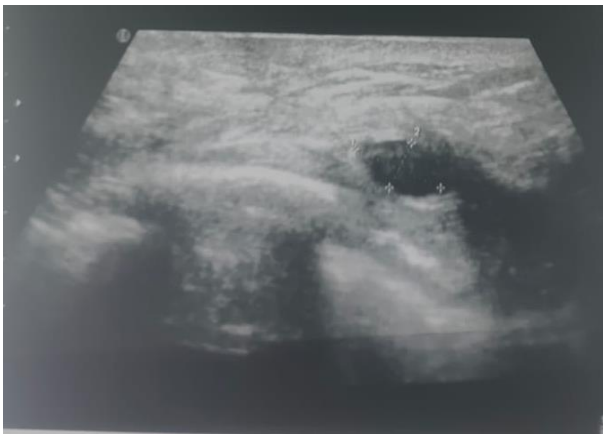

Image 1: Breast ultrasound showing a subcutaneous, well-defined oval lesion with a tissular echo structure, measuring 32 x 18 mm

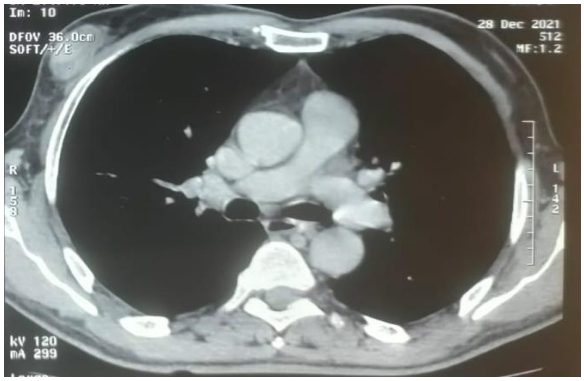

Image 2: The thoraco-abdomino-pelvic CT scan confirming the presence of a right breast nodule

CASE 2

This concerns a 62-year-old woman, G3P3, postmenopausal, hypertensive for 15 years, who incidentally discovered a left breast nodule during a cardiology consultation. Upon the initial examination, the masse is located at the junction of the lower quadrants of the left breast, measuring 7 cm in diameter, without axillary lymphadenopathy. Mammography revealed a large nodular formation with polylobed contours, containing some microcalcifications in the lower quadrants of the left breast (Image 3). The complementary ultrasound describes a lesion process on the 6 o'clock line of the left breast, measuring 90/65/60 mm, containing cystic areas with polylobed contours classified as BIRADS 5. After a wide tumorectomy, the histological study of various samples taken from the tumor focused on mammary tissue infiltrated by a well-defined peripheral carcinomatous tumor proliferation, predominantly of a solid architecture (estimated at 85%). It consists of masses of cells with relatively monomorphic nuclei, finely granular chromatin, and scanty eosinophilic cytoplasm. These masses are traversed by small cords of fibro-vascular stroma. Mitotic figures are rare, estimated at 4 mitoses per 10 fields. Additionally, focal areas, estimated at 15%, of encapsulated papillary carcinoma were observed (anti-p63 antibody: negative). There was no infiltrating component in all examined samples and no vascular emboli. The lateral resection margins are clear, with the closest being the inner margin, located 5mm away. The tumor showed positive hormone receptor expression in 90% of cells and a Ki67 proliferation index of 15%. Immunostains for p63, chromogranin A, and synaptophysin were negative. Adjuvant radiotherapy was scheduled. During the dosimetric CT scan, two tissue masses centered on K3 and K9 were observed. A CT-guided biopsy of the costal mass confirmed a bony localization of mammary adenocarcinoma, the immunohistochemical study showed the following: Anti-CK20 antibody: Negative, Anti-estrogen receptor antibody: Intense staining in 100% of tumor cells, Anti-progesterone receptor antibody: Displays moderate to intense staining in 90% of tumor cells, Herceptest negative: Indicates weak and incomplete staining .A PET FDG scan, as part of the overall staging, revealed pathologically active lymph node involvement in the mediastinum and several active secondary bone lesions (Image 4). The patient is receiving a combination of an aromatase inhibitor, a CDK4/6 inhibitor, and zoledronic acid. She is tolerating the treatment well, and there is no evidence of disease progression.

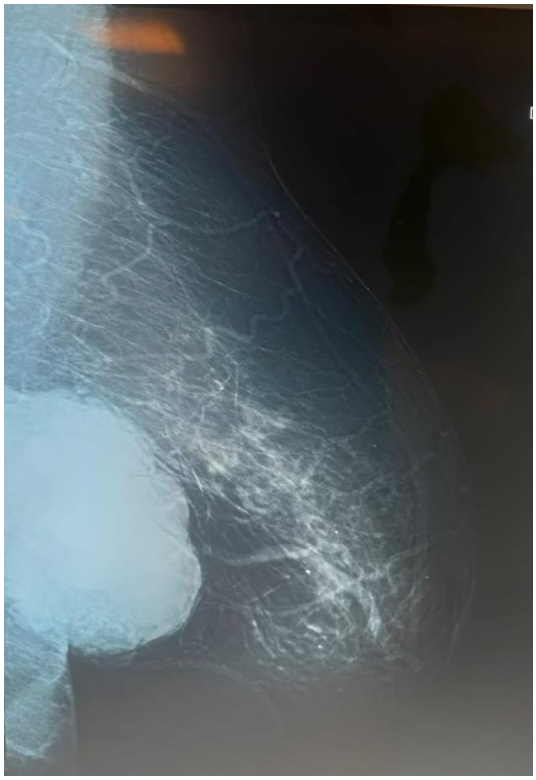

Image 3: Mammography revealing a large nodular formation with polylobed contours, containing some microcalcifications in the lower quadrants of the left breast

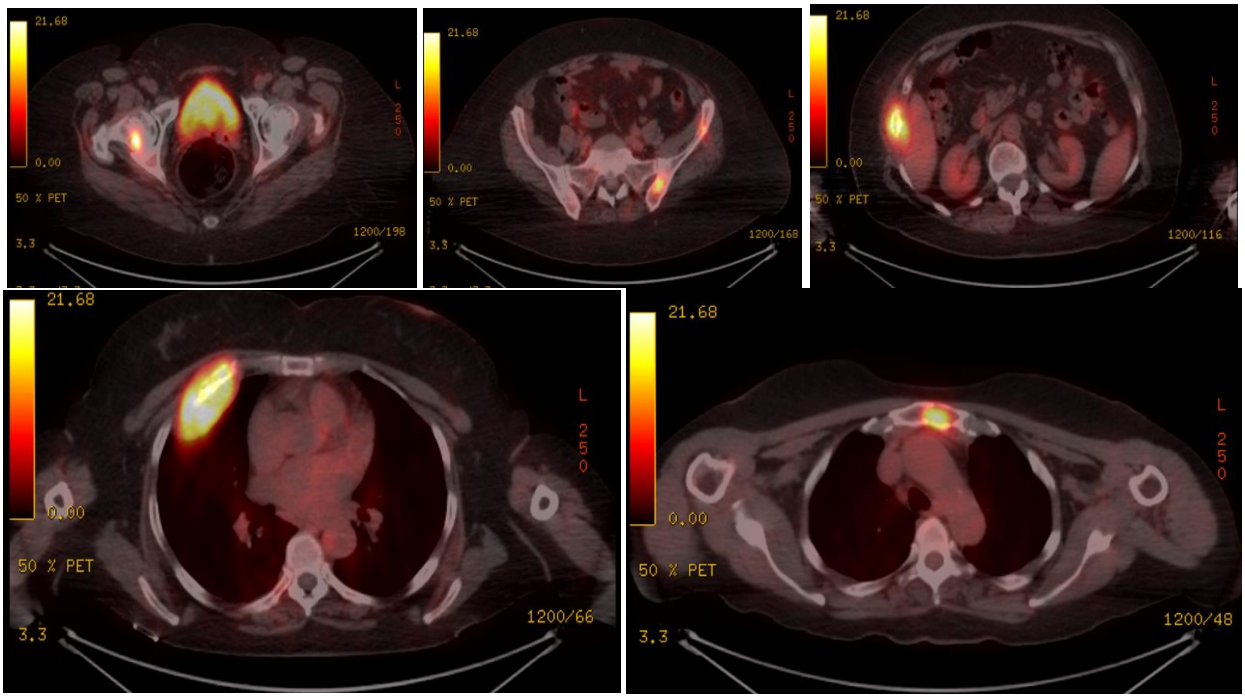

Image 4: PET FDG scan, revealing pathologically active lymph node involvement in the mediastinum and several active secondary bone lesions

DISCUSSION

Encapsulated papillary carcinoma (EPC) of the breast, a rare occurrence accounting for only 0.5-1% of all malignant cases worldwide [3], is considered a transitional stage between ductal carcinoma in situ (DCIS) and invasive carcinoma [4]. Based on histological features, EPC can be categorized into three subtypes: Pure EPC, EPC associated with DCIS, and EPC associated with invasive carcinoma [5].

EPC is commonly thought to affect elderly women; however, it can occur across all age groups. While the majority of cases are found in women, instances have been diagnosed in men as well. In a comprehensive study conducted by Gbowsky *et al*., [6] a total of 917 cases of EPC were identified in the California Registry. The median age of patients with EPC was 69.5 years (ranging from 27 to 99 years). The vast majority of cases were diagnosed in women, accounting for 96.5% while 3.5% of cases occurred in men. About 47% of cases were categorized as carcinoma in situ (CIS), and the remaining 53% were classified as invasive. Among the invasive cases, an overwhelming majority were localized at the time of diagnosis (89.6%; n=439), with 7.8% classified as regional disease, demonstrating either direct extension into adjacent tissue or axillary lymph node involvement. Distant-stage cases at the time of diagnosis were exceptionally rare, accounting for only 0.4%, with a reported total of 2 cases.

This type of carcinoma typically manifests as a well-defined central and subareolar round mass. Specific imaging features distinguishing encapsulated papillary carcinoma from other papillary lesions are lacking. Nevertheless, encapsulated papillary carcinomas tend to present as larger masses, often exhibiting a cystic growth pattern [7].

From a histological standpoint EPC is characterized by fine fibrovascular stalks covered by neoplastic epithelial cells of low or intermediate nuclear grade, it is typically found within a cystic space and enclosed by a fibrous capsule. Notably, there are usually no myoepithelial cells along the papillae or at the periphery of the lesion. Given the absence of myoepithelial cells and the rarity of axillary lymph node metastases, it is generally considered a self-contained indolent invasive carcinoma with a prognosis similar to that of carcinoma in situ [8], at the same time due to the lack of myoepithelial cells, it has been described that a subset of EPC cases has invasive potential and are able to develop local and/or distant metastases [9].

No established evidence-based guidelines currently guide the management of encapsulated papillary carcinoma (EPC) due to its infrequent occurrence. In summary, if there is no invasion beyond the fibrous capsule, encapsulated papillary carcinoma should be graded based on its nuclear grade and staged as "pTis (DCIS)" to avoid overtreatment. In cases of frank invasion, the Nottingham grade and tumor stage should be determined solely based on the morphological features and pathological size of the invasive component, and HR and HER status should be assessed on the invasive component exclusively.

Primary treatment, following EPC diagnosis post-biopsy, involves complete surgical excision, whether through breast-conserving surgery or

mastectomy. The omission of Sentinel Lymph Node Biopsy (SLNB) in cases where pure encapsulated papillary carcinoma (EPC) is still a matter of debate. Some researchers, however, advocate for SLNB as a viable surgical option when invasive EPC is identified, citing its potential benefits in both prognosis and treatment [10]. Beyond surgical excision, the efficacy of adjuvant treatments such as radiotherapy, chemotherapy, endocrine therapy, and HER2-targeted therapy in encapsulated papillary carcinoma (EPC) is not well-established. The decision for adjuvant treatment in EPC should ideally consider the malignant potential of the invasive tumor cells rather than the in-situ components. Consequently, prior publications have suggested adjuvant radiotherapy, chemotherapy, and endocrine therapy for patients with EPC associated with invasive carcinoma [11].

With appropriate local therapy, encapsulated papillary carcinoma, in the absence of associated infiltrating carcinoma, carries a very favorable prognosis. Although rare, instances of lymph node metastases have been reported, often revealing typical papillary features. The presence of associated ductal carcinoma in situ in adjacent breast tissue increases the risk of local recurrence. Therefore, complete surgical excision of the lesion, along with extensive sampling of the surrounding breast tissue, is crucial for treatment and assessing the risk of local recurrence. Lesions presenting with a growth pattern similar to encapsulated papillary carcinoma but with nuclear pleomorphism, increased mitotic activity, and/or a triple-negative or HER2-positive phenotype should be treated and managed as invasive breast cancer.

CONCLUSION

In summary, encapsulated papillary carcinoma, commonly occurring in elderly women, generally exhibits a favorable prognosis. Despite its classification as a malignant tumor in situ with indolent behavior, the absence of myoepithelial cells suggests potential for metastasis in EPC.

Competing interests: We (authors) declare that we have no conflict of interest.

Authors' Contribution: Mouhcine Hommadi and Khalid hadadi contributed equally to the work and should be considered co-first authors, Maroua Benlemlih, Elamin Marnouch, Abdelhak Maghouss, Amine Bazzine, Noha zaghba, Issam Lalya, Khalid Andaloussi Saghir, Mohammed Elmarjany, and Hassan Sifat designed and coordinated research and drafted the manuscript. All authors read and approved the final manuscript.

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