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Case Report

Mammary type adenocarcinoma of the vulva: a rare case report

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ABSTRACT

A primary vulvar mammary type of adenocarcinoma is an exceptionally rare malignancy that is thought to arise from mammary-like vulvar glands. Its low incidence is partly responsible for the lack of guidelines for patient management. We present a case of a 74-year-old woman who presented with a palpable mass in her vulvar region and was submitted to an excisional biopsy, which was diagnosed as mammary-type adenocarcinoma. The patient then underwent a partial vulvectomy with unilateral inguinal sentinel lymph node biopsy followed by adjuvant hormonal therapy. This case report highlights the challenges in diagnosing and managing this rare entity and emphasizes the importance of a multidisciplinary approach to providing optimal care for patients with a mammary type of adenocarcinoma of the vulva.

Keywords: Mammary type adenocarcinoma, Rare vulval cancer, Ectopic breast tissue

INTRODUCTION

Vulvar cancers represent 5% of all gynecologic malignancies and only 10% of vulvar cancers are adenocarcinomas. Adenocarcinomas of the vulva include Bartholin gland carcinomas, extramammary Paget disease, sweat gland carcinoma, mammary-like adenocarcinomas, and adenocarcinomas of metastatic origin. To our knowledge, there have been 36 reported cases of primary breast carcinoma of the vulva until 2017 since the first documented case by Greene in 1935. Due to its rarity, there is limited information available regarding its clinical behaviour, optimal management, and prognosis.[1] Here, we present a case of mammary-type adenocarcinoma of the vulva in a 74-year-old woman, with a focus on the diagnostic challenges, treatment approach, and patient outcome.

CASE REPORT

A 74-year-old diabetic and hypertensive postmenopausal woman, Eastern cooperative oncology group-performance status (ECOG-PS) grade 0, without a significant personal or family history of cancer disease, presented with a vulvar mass on the right side for the last 1 year. On physical examination, a 3-cm firm, non-tender mass was noted in the right labia majora. The overlying skin appeared intact, with no signs of inflammation or ulceration. The remainder of the pelvic examination was unremarkable, and there were no palpable inguinal lymph nodes.

Given the atypical presentation, the patient underwent a biopsy of the vulvar mass. Histopathological examination revealed the presence of a well-differentiated mucinous adenocarcinoma (Figure 1). Immunohistochemical (IHC) staining demonstrated positivity for carcinoembryonic antigen (CEA), GATA 3, CK7, estrogen receptor (ER), and progesterone receptor (PR), consistent with mucinous adenocarcinoma (Figure 2). Further workup including pelvic magnetic resonance imaging (MRI) and positron emission tomography-computed tomography (PET-CT) revealed a mild thickening on the right side of the vulva secondary to post-intervention inflammatory change without evidence of distant metastasis. Clinical examination, breast ultrasound, and mammography did not reveal any signs of a primary breast tumor.

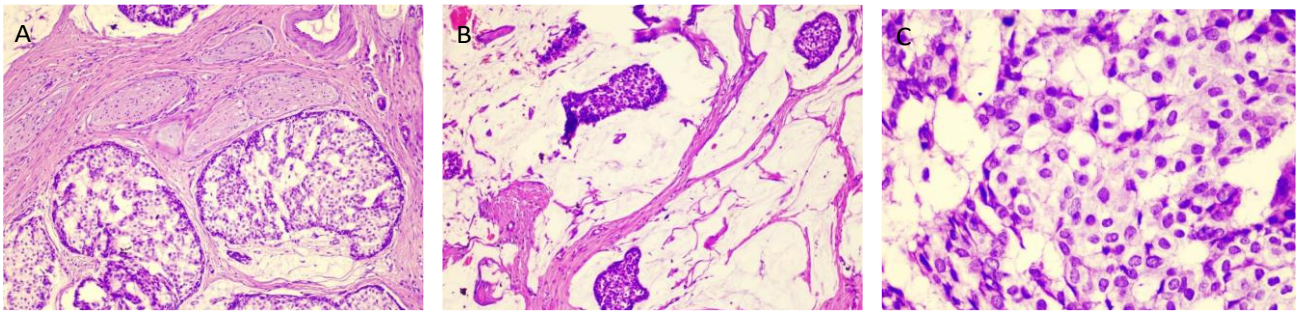

Figure 1: A) Low power photomicrograph of the tumor showing cellular areas with cells arranged in cribriforming pattern (H&E, 10x). B) Another low power photomicrograph showing a relatively paucicellular area with tumor cell morules floating in pools of abundant extracellular mucin (H&E, 10x). C) High power photomicrograph showing a relatively monomorphic population of tumor cells with round regular nuclei and abundant clear cytoplasm (H&E,40x).

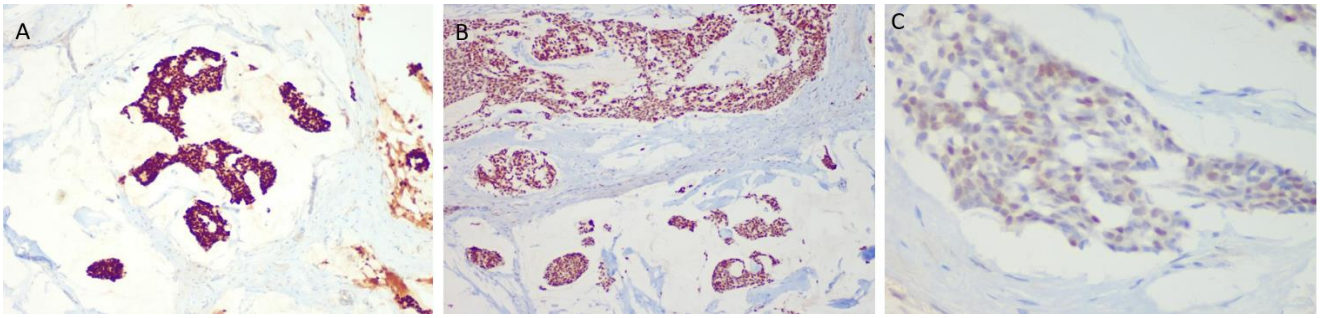

Figure 2: A) Tumor cells showing diffuse strong nuclear expression of GATA3 immunostain (IHC:GATA3, 10x). B) Tumor cells showing diffuse strong nuclear expression of ER (IHC:ER, 10x). C) Tumor cells showing patchy and weak nuclear expression of PR (IHC:PR, 40x).

Subsequently, the patient underwent radical partial vulvectomy with unilateral inguinal sentinel lymph node dissection. Surgical pathology confirmed the diagnosis of well-differentiated mammary-type mucinous adenocarcinoma, with negative surgical margins and no lymphovascular invasion. The right inguinal lymph node was negative for malignancy. Given the ER and PR positivity on IHC, the patient was referred to medical oncology for consideration of adjuvant hormonal therapy. She was initiated on adjuvant letrozole and her postoperative recovery was uneventful. Regular follow-up visits including history, physical examination, and pelvic imaging were planned to monitor for disease recurrence.

At the three-month follow-up, the patient remained asymptomatic with no evidence of disease recurrence on clinical examination and imaging studies. She is being closely monitored by a multidisciplinary team including gynecologic oncology, medical oncology, and radiation oncology.

DISCUSSION

About the aetiopathogenesis of mammary-type vulval adenocarcinoma, different hypothesis is mentioned in the literature. It is believed that they originate from ectopic mammary tissue, which arises as a result of incomplete involution of the ectodermal mammary ridges. The mammary ridges develop from the fourth week after conception during embryogenesis and usually regress thereafter, except in the thoracic region where the mammary glands complete their development.[1] These tumors are believed to be derived from anogenital mammary-like glands located in the interlabial folds, which have subtle histological and ultrastructural differences from breast tissue. This ectopic breast tissue is similar to orthotopic breast tissue in being hormone sensitive and can also undergo benign or malignant changes.[2,3] Thus, diagnosis of this malignancy poses significant challenges due to its histological similarity to breast carcinoma. Since it arises from mammary-like tissue within the vulva, the possibility of metastatic breast carcinoma involving the vulva must always be considered and excluded through comprehensive IHC analysis and clinical correlation. Any of the various histologic subtypes of breast cancer including infiltrating ductal, lobular, mucinous, and mixed ductal and lobular carcinoma, can also occur similarly in the vulva.[4,5] To differentiate between primary and metastatic vulval cancer following criteria should be met: a morphologic pattern consistent with breast carcinoma; the presence of estrogen and progesterone receptors; and/or and positivity for breast cancer markers such as epithelial membrane antigen, carcinoembryonic antigen, and glandular keratins. The most common clinical presentation was a painless, solitary nodule, arising most often in the labia majora which was

consistent with our case. These are locally aggressive tumors and lymph node metastases in present in approximately 60% of cases.[6,7]

The optimal management of mammary-type adenocarcinoma of the vulva remains uncertain due to the scarcity of cases. Treatments are often extrapolated from guidelines for orthotopic breast cancers due to their resemblance to breast tissue. Surgical excision with negative margins and lymph node assessment is the mainstay of treatment for localized disease. Surgical procedures include radical vulvectomy or radical local excision and lymphatic involvement may be assessed by sentinel lymph node biopsy (SLNB) or uni/bilateral lymphadenectomy.[8] However, the role of adjuvant therapy, including radiation therapy (RT), chemotherapy, and HT, is not well defined and varies according to the IHC study and disease staging. Given the ER positivity observed in some reported cases, adjuvant HT may be considered in managing mammary-type adenocarcinoma of the vulva.

In advanced stages, neoadjuvant therapy followed by surgery and/or adjuvant postoperative chemoradiation has been suggested. The role of gene-targeted or immunotherapy approaches remains to be evaluated.[9] The addition of CDK4/6 inhibitor agents to hormonal therapy can result in significant improvement in progression-free survival. Trastuzumab (Herceptin) can be used in HER2-positive cases, but its use has been reported in very few cases in the literature.[10]

Despite the lack of evidence-based guidelines, this treatment approach was based on a multidisciplinary consensus to minimize the risk of disease recurrence. The patient's favourable response to treatment and absence of disease recurrence at the three-month follow-up underscore the potential for successful outcomes with a multimodal treatment approach.

Close surveillance following treatment is essential, as the natural history and long-term outcomes of this malignancy are not well understood. There is a need to develop well-established guidelines for the treatment of adenocarcinoma of mammary type with primary manifestation in the vulva. Reporting these clinical cases will increase the knowledge about the pathogenesis of these rare lesions and help to debate their clinical management and outcome.

CONCLUSION

By reporting these rare cases, we intend to contribute to the limited body of literature on this entity's clinical presentation, diagnosis, and management. A multidisciplinary approach is essential for providing individualized and optimal care for patients with mammary-type adenocarcinoma of the vulva. Further research and collaboration are warranted to understand better the natural history, optimal treatment strategies, and long-term outcomes of this rare vulvar malignancy.

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