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Adenocarcinoma of Mammary-Like Glands of the Vulva: A Case Report and Review of Literature

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Introduction

An estimated 4% of gynecologic cancers are vulvar carcinomas. Squamous cell carcinomas make up around 90% of vulvar malignancies, while melanomas make up about 5 %. [1,2] Adenocarcinomas, transitional cells, adenoid cystic, and adenosquamous carcinomas are among the others. Adenocarcinoma of the vulva of the mammary gland type (AMGT) has a poorly understood aetiopathogenesis. It is still unclear from where the mammary-like vulvar glands came and how they became the source of a rare kind of primary vulvar cancer. They were once thought to originate from ectopic mammary tissue as a result of insufficient involution of the mammary ridges.

Van der Putte and van Gorp, however, have recently hypothesised that these tumours develop from interlabial glands that resemble anogenital mammary glands.[3,4]

The mammary-like vulvar gland tissue is hormone sensitive and able to undergo benign or malignant changes, just like orthotopic breast tissue. Their malignant transformation is extremely rare.[5,6] Patients may remain asymptomatic or experience symptoms as a result of the physiological changes that are common during menarche, pregnancy, or lactation. As a differential diagnosis, one should include adenocarcinomas from Bartholin glands, extramammary Paget disease, metastatic adenocarcinomas, and sweat gland carcinoma. Yet whenever a vulvar tumour is found, the possibility of AMGT should be considered, especially if a histological examination reveals breast tissue morphology or a history of breast cancer.

The first case of a vulvar mammary-type cancer was documented by Greene in 1936.[7] The lack of patient management guidelines is a result of the low occurrence, only 36 cases of adenocarcinoma of the vulva (AMGT) had been documented up until 2017.[8] Additionally because the aetiopathogenesis of these tumours is poorly understood, there is no approved classification scheme for these tumours.[9] Treatments are commonly extrapolated from those for orthotopic breast tumours nevertheless, the optimum surgical margins and the staging of the lymph nodes are still up for debate.

Here, we report a case report of a patient who was diagnosed with AMGT of the vulva and how she was managed. The patient's consent was taken. No ethical concerns were brought up, and confidentiality was maintained.

Case Presentation

Our patient, a 47-year-old P2L2 female, came to the oncology OPD with a vulva biopsy report that was suggestive of a vulva malignancy. She had multiple nodules involving bilateral labia majors with complaints of intense itching and occasional bleeding from the nodules. In addition, she gave a history that it started as a solitary nodule over the labia major one year back. Our pathology department reviewed the histology slides with IHC markers, and a diagnosis of adenocarcinoma of mammary gland type of vulva with positive P40, CK7, ER, GATA-3 and EPCAM focal was given.S100, HMB45, P63, UROLAKIN, and CK20 were determined to be negative

The whole body PET scan showed several avid moderately enlarged lymph nodes in the bilateral inguinal and external iliac regions, as well as numerous avid nodular lesions in the vulva including the bilateral labia minor and major, mostly along the midline. Metastatic deposits were confirmed by FNAC from the right inguinal lymph nodes, indicating stage IIIB. The patient was counselled regarding the aggressive nature of the disease, the prognosis, and the chances of recurrence and relapse.

After a multidisciplinary discussion with the tumour board, it was decided to offer the patient the option of new adjuvant chemotherapy (NACT) followed by surgery and radiotherapy. Four cycles of NACT with carboplatin and paclitaxel were given to the patient.Repeat PET Scan after one month of the last cycle revealed heterogeneously enhancing nodular thickening with FDG avidity in the vulva involving bilateral labia minor and major, predominately along the midline (reduced in extent and avidity).

Non-FDG avid centimetric sized bilateral inguinal lymph node (reduced in size and avidity). Non-FDG avid very small bilateral external iliac lymph node (reduced in size and avidity). Overall scan findings were suggestive of a favourable response to treatment.

On examination, no vulval nodules were seen, and only diffuse oedema involving bilateral labia majors was there (image 1). No palpable inguinal lymph nodes were found. MRI pelvis was done and its findings were reported as mild thickening of both sides of the vulva, left>right with mild soft tissue oedema involving the clitoris and mons pubis. She was optimized for surgery and a radical vulvectomy with bilateral inguinal and bilateral pelvic lymph node dissection was done (images 2-4). The final postoperative histopathological report showed residual adenocarcinoma with no perineurial or lymphovascular invasion. All dissected 41 bilateral inguinofemoral and pelvic lymph nodes were found free of disease. The patient recovered well and all the wounds were healthy on follow-ups. After further discussion in the tumour board plan for postop radiotherapy (50Gy in 25 fractions to PTV Primary and

45Gy/25frs to PTV pelvis along with gap correction) with weekly concurrent cisplatin was offered to the patient. The patient is currently on a routine monthly follow-up and doing well.

IMAGE 1

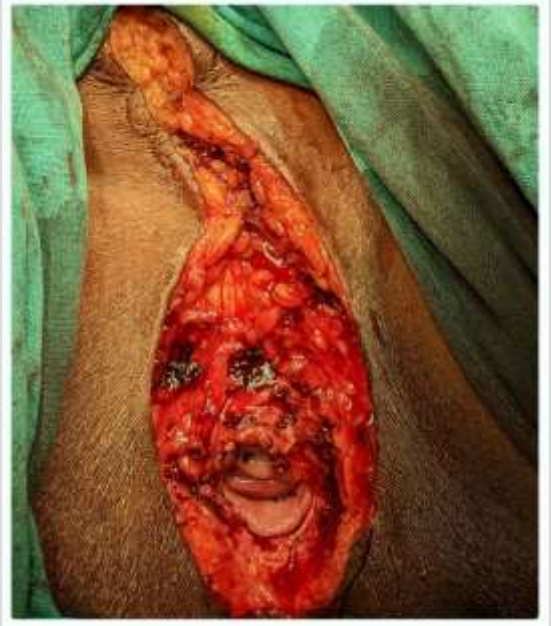

IMAGE 2

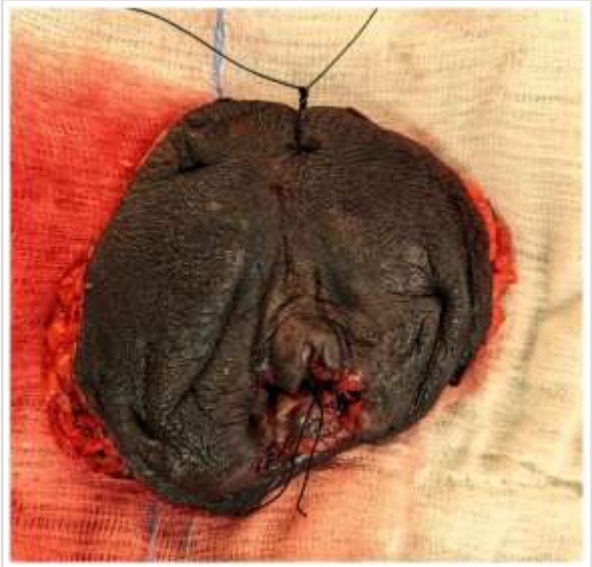

IMAGE 3

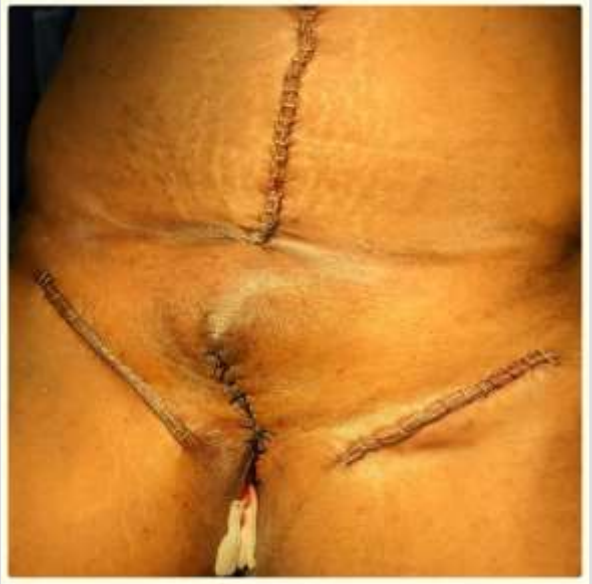

IMAGE 4

Discussion

Vulvar adenocarcinomas are rare, and those that come from tissue that resembles the vulvar mammary gland are extremely rarer. While Greene published the first instance of a mammary-like tumour of the vulva in 1936, Hartung initially recorded the presence of a completely formed mammary gland in the left labium majus in an elderly woman in her 80s in 1872. [5-7] Patients with vulvar mammary-like adenocarcinomas are 62.5 years old on average. [8] With a mean age at diagnosis of about 67 years (range 47-87), and half of those older than 80 years.[10-13] Recently described data suggest that this disease mostly affects older women however our patient was younger than usual. Overall, the vulvar lesion is described as an asymptomatic solitary nodule, mostly located on the labia majora. This description is coincident with our case as the patient first reported the development of a single nodule however, over the period of one year number of nodules increased and involved bilateral labia majors.

An incisional biopsy is crucial in establishing the diagnosis, as the list of differential diagnoses of a vulvar lump is long, ranging from benign entities such as Bartholin abscess to vulvar mesenchymal neoplasm and malignant tumours such as squamous cell carcinoma. For the diagnosis of vulvar carcinoma of mammary gland type, histopathological patterns are essential.[12,14] Diagnostic criteria include (1) morphology consistent with breast carcinoma, (2) positive ER and/or PR, (3) positivity for typical immunohistochemical breast markers and (4) the presence of carcinoma in situ or non-neoplastic breast tissue, adjacent to the tumour.[15,16] Furthermore, it is necessary to exclude metastasis disease from orthotopic breast carcinoma or other organs.[16] Three out of these four criteria were met in our case and the metastatic origin was excluded. There was no carcinoma in situ or non-neoplastic breast tissue present in ad- jacent areas. The patient had no lesion on the breast and the mammogram was normal. The presence of GATA3-positive in the vulvar tumour bed confirmed mammary- like glands origin.

Because of the resemblance between this entity and breast carcinoma, using the orthotopic breast cancer treatment approaches is typically advised.[9,15] Nowadays, neoadjuvant chemotherapy is used to downstage big tumours so that breast conserving therapy can be offered to patients with locally advanced breast cancer.[17] Extrapolating from the evidence of the beneficial role of NACT in breast cancer similar treatment approach was taken in our case. This is by far the first case reported in the literature of metastatic adenocarcinoma of vulva mammary gland type treated with Neo adjuvant chemotherapy. PET CT after completion of NACT showed a favourable response with the dissolution of vulval nodules and reduction in size and FDG avidity of inguinal and pelvic lymph nodes.

Guidelines for the care of primary adenocarcinomas of vulvar glands that resemble the mammary glands cannot be established due to the lack of reported cases of this condition. Surgical treatment has been adopted in most cases, from local excision to radical vulvectomy with bilateral inguinofemoral lymphadenectomy.[18-19] Inguinofemoral lymphadenectomy and pelvic lymphadenectomy, if necessary, should be included in the surgical treatment since the pathologic state of the lymph nodes is the most significant prognostic factor in vulvar cancer. We performed a pelvic lymphadenectomy after identifying the pelvic nodal involvement in our patient's PET CT scan. As far as we are aware, this is the second instance in which a patient with vulvar cancer that resembled a mammary tumour underwent bilateral pelvic lymphadenectomy surgery in addition to inguinofemoral lymphadenectomy. Virginia Benito et al. reported the first instance in 2013.[20]

Since AMGT is a rare disorder, there aren't any established recommendations, by presenting this case, we hope to advance understanding of the pathophysiology of these uncommon lesions through histological and IHC studies as well as stimulate discussion about their clinical treatment and prognosis. We do, however, consider that care and follow-up should be the same as for orthotopic breast cancer of a similar stage given the parallels and biological behaviour of both diseases.

Conclusion

There are various hypotheses about the aetiopathogenesis of adenocarcinoma of mammary gland type (AMGT) of the vulva in the literature. Despite responding to the same physiological and pathological changes as the breast, vulvar mammary like tissue seldom develops into a mammary gland cancer, and there aren't many examples of AMGT of the vulva documented in the literature. Treatment is still debatable, although the same strategy that is employed for orthotopic breast cancer at a comparable stage could be applied. In summary, this is the first article to describe a case of metastatic primary adenocarcinoma of mammary like glands of the vulva successfully treated with a combination of NACT with surgery. By reporting these cases, we intend to increase the knowledge about the pathogenesis and clinical management of these lesions.

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