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Case Report

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Proximal Type" Epithelioid Sarcoma of the Vulva: A Case Report

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ABSTRACT

Epithelioid sarcoma (ES) is a rare malignant soft tissue tumor, with the proximal-type variant (PES) representing an uncommon and aggressive subtype. Vulvar sarcoma predominantly affects young women, with a mean age of approximately 38 years. We report a case of a 22-year-old G1L1 Asian woman presenting with a painless, solid mass in the right labia majora, without tenderness or surface ulceration. Surgical management consisted of a right hemi-vulvectomy with a 2 cm margin from the tumor. Histopathological evaluation confirmed the diagnosis of epithelioid sarcoma of the vulva, and the surgical margins were free of tumor involvement.

Postoperatively, the patient was followed every three months for the first two years and annually thereafter. Over a ten-year follow-up period, no evidence of local recurrence or metastasis was observed, and the patient experienced two successful pregnancies during this time. This case highlights the importance of considering vulvar sarcomas in the differential diagnosis of nonspecific vulvar lesions to ensure early recognition, accurate diagnosis, and appropriate management.

Introduction

Epithelioid sarcoma (ES), first described by Enzinger in 1970, is a rare malignant soft tissue neoplasm characterized by both epithelial and mesenchymal differentiation [1]. Based on its anatomical distribution and clinical behavior, ES is classified into two distinct subtypes: distal-type, which typically arises in the extremities, and proximal-type, which primarily occurs in the trunk, pelvis, and pubic regions, often exhibiting a more aggressive course and poorer prognosis [2]. Proximal-type epithelioid sarcoma (PES) is an uncommon and highly malignant variant that tends to affect young adults [3]. Due to its rarity, published cases remain limited, and its clinical and pathological characteristics are not yet fully understood. Clinically, PES of the vulva may mimic vulvar squamous cell carcinoma (SCC) or present as a benign-appearing

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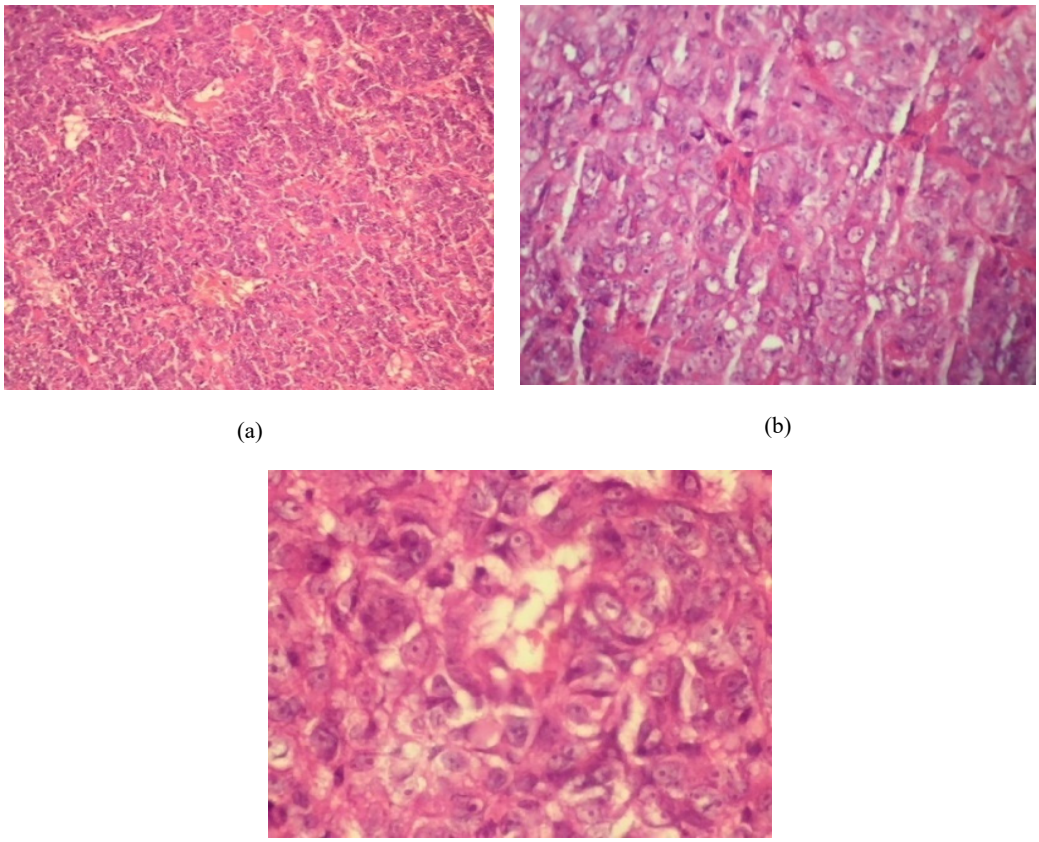
mass with well-defined margins, leading to potential diagnostic delays. Histologic grading remains the most significant prognostic indicator, as undifferentiated tumors exhibit rapid proliferation and early metastasis, while well-differentiated lesions may be amenable to local excision. Currently, there is no universally accepted treatment guideline for epithelioid sarcoma of the vulva, as it is a rare condition. However, wide local excision with adequate surgical margins is generally considered the primary treatment of choice [4-6]. In this report, we present a rare case of proximal-type epithelioid sarcoma of the vulva in a 22-year-old woman who underwent successful surgical management.

Case Presentation

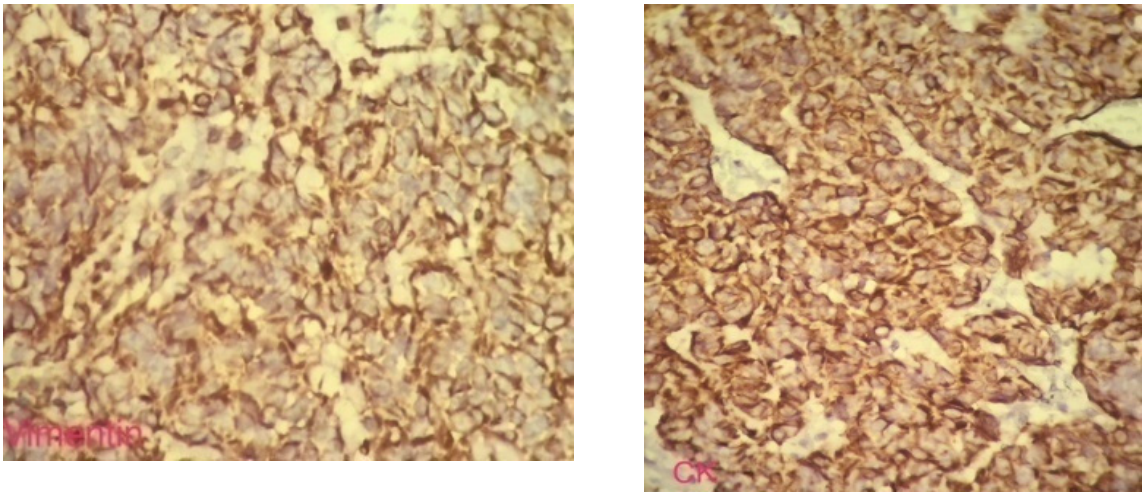
A 22-year-old G1L1 Asian woman with a body mass index (BMI) of 24 presented with a gradually enlarging, painless mass in the upper portion of the right labia majora over three months. On physical examination, a firm, mobile, non-tender, and well-circumscribed mass measuring approximately 4 cm in diameter was palpated, extending toward the mons pubis without adherence to underlying bone or evidence of surface ulceration. Preoperative pelvic ultrasonography demonstrated a hypoechoic mass lesion measuring 40 × 30 mm located in the upper right labia majora, approximately 23 mm beneath the vulvar skin surface. Chest and abdominopelvic computed tomography (CT) scans revealed no signs of metastatic disease. Routine laboratory investigations and Pap smear results were within normal limits. The patient underwent a right hemi-vulvectomy with a 2 cm surgical margin from the gross tumor border. The postoperative course was uneventful, and she was discharged two days after surgery in good general condition. Histopathological examination confirmed the diagnosis of epithelioid sarcoma of the vulva, with tumor-free surgical margins. Microscopic evaluation revealed sheets of mitotically active polygonal cells with a high nuclear-to-cytoplasmic ratio, abundant eosinophilic cytoplasm, large eccentric nuclei, and prominent nucleoli. Collagen bundles and mucinous material were evident in the stroma (Figure 1).

Microscopic evaluation revealed sheets of mitotically active polygonal cells with abundant eosinophilic



(a) (b) (c)

Fig. 1. Epithelioid sarcoma: tumor is composed of sheets of polygonal cells with high nuclear to cytoplasmic ratio, abundant eosinophilic cytoplasm, large eccentric nuclei, and prominent nucleoli. *Hematoxylin & Eosin staining*; ×40 (a), ×100 (b), and ×400 (c) magnification.



(a) (b)

Fig. 2. Immunohistochemical staining showing positivity of vimentin (a) and CK (b).

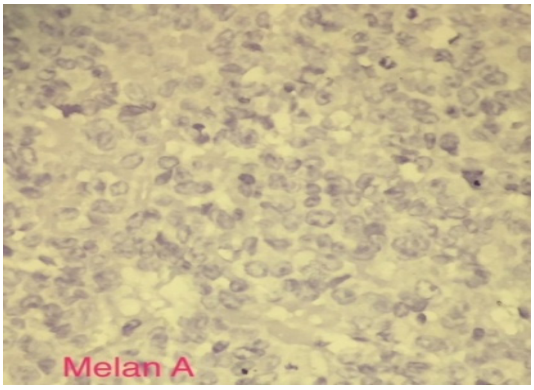


Fig. 3. Immunohistochemical staining showing negative Melan-A.

cytoplasm, high nuclear-to-cytoplasmic ratio, large eccentric nuclei, and prominent nucleoli. The cells were embedded within a collagenous stroma, with focal areas of mucinous material observed (Figure 1). Histopathological features were consistent with proximal-type epithelioid sarcoma. Immunohistochemical analysis demonstrated strong and diffuse positivity for vimentin, cytokeratin (CK), and CD117 (Figure 2), confirming the dual epithelial–mesenchymal differentiation characteristic of this tumor type. In contrast, Melan-A, leukocyte common antigen (LCA), CD20, CD3, actin, desmin, and alpha-fetoprotein (AFP) were negative (Figure 3), effectively excluding morphologically similar neoplasms such as melanoma, lymphoma, and myogenic tumors. The Ki-67 proliferative index was approximately 10% (Figure 4), indicating relatively low proliferative activity, in line with the indolent clinical behavior often observed in these cases. The diagnosis of proximal-type epithelioid sarcoma was established based on the combination of anatomical location, morphological characteristics, and the immunophenotypic profile. Following surgery, the patient was followed up every three months for the first two years and annually thereafter. During the ten-year follow-up period, she remained disease-free with no evidence of local recurrence or distant metastasis. Notably, she achieved two successful pregnancies during this interval.

Discussion

We report a rare case of proximal-type epithelioid sarcoma (PES) of the vulva in a 22-year-old woman who presented with a painless, mobile mass in the right labia majora. The most common initial manifestation of vulvar epithelioid sarcoma (ES) is typically a slowly enlarging, painless subcutaneous nodule, often mimicking a benign lesion and thereby contributing to diagnostic delays [7]. Due to its deceptively benign clinical appearance, ES

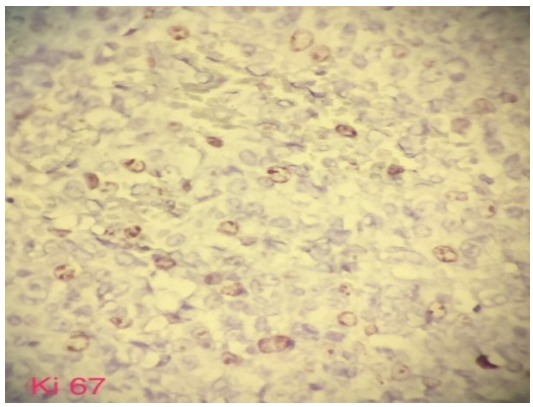


Fig. 4. Ki67 immunohistochemical staining: tumor proliferative index is about 10%.

may be misdiagnosed and inappropriately managed with limited excision or biopsy. In a twenty-year retrospective study at Johns Hopkins University, seven cases of vulvar sarcoma were identified, among which only one was diagnosed as epithelioid sarcoma; this patient underwent radical vulvectomy without adjuvant therapy and experienced no recurrence [8]. Similarly, Dash et al. reported four cases of vulvar epithelioid sarcoma, one of which presented with pulmonary metastasis and received palliative chemotherapy, while the remaining three remained disease-free for over five years after surgical treatment [9]. To date, the optimal management strategy for PES of the vulva remains undefined, largely due to its rarity and the paucity of prospective studies. Wide local excision or radical vulvectomy with clear surgical margins of at least 2 cm is generally recommended as the primary treatment approach [10]. The role of adjuvant therapy, including radiotherapy and chemotherapy, remains controversial. Adjuvant radiotherapy may be considered for high-grade tumors, close or positive margins, or recurrent disease; however, its impact on overall survival has not been conclusively demonstrated. Likewise, chemotherapy has shown limited efficacy, primarily reserved for metastatic or recurrent cases, with anthracycline-based regimens (such as doxorubicin) being the most commonly employed agents [11-13]. Our patient underwent right hemi-vulvectomy without adjuvant therapy and has remained disease-free for ten years postoperatively, highlighting the potential for excellent long-term outcomes with complete surgical resection and negative margins. Given the scarcity of cases, management should ideally involve a multidisciplinary team, including gynecologic oncologists, pathologists, and radiologists, to ensure accurate diagnosis and individualized treatment planning.

Conclusion

In conclusion, proximal-type epithelioid sarcoma of the vulva is an exceptionally rare malignancy with variable clinical behavior and uncertain therapeutic guidelines. Clinicians should maintain a high index of suspicion for vulvar sarcomas when evaluating nonspecific vulvar masses, as early recognition and adequate surgical excision are essential to achieving favorable outcomes.

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Ethical Considerations

Key Clinical Message

Proximal-type epithelioid sarcoma of the vulva is an exceptionally rare malignancy that can clinically mimic benign lesions. Maintaining diagnostic vigilance and performing early histopathological evaluation are crucial. Complete surgical excision with negative margins remains the cornerstone of treatment, offering excellent long-term outcomes and minimizing the risk of recurrence or metastasis.

Author Contributions

F.B. contributed to the investigation, original draft preparation, and manuscript review and editing. P.R. participated in the investigation, data interpretation, and manuscript writing and revision.

Z.SH. and M.N. were responsible for data curation, resource collection, and manuscript preparation. A.Z. contributed to study conceptualization, methodology design, project administration, supervision, and final manuscript validation. All authors have read and approved the final version of the manuscript.

Ethics Statement

This study was conducted in accordance with the principles of the Declaration of Helsinki. The research protocol was reviewed and approved by the Ethics Committee of Isfahan University of Medical Sciences (approval code: IR.ARI.MUI.REC.1403.229).

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Conflict of Interests

The authors have no conflict of interest to declare.

Consent

Written informed consent has been obtained from the patient involved in this study.

Data Availability Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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