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Correspondence

Proximal-type epithelioid sarcoma of the vulva initially misdiagnosed as Bartholin cyst: A case report

Rafael Everton Assunção Ribeiro da Costa [a,*], Luis Felipe Rodrigues Brandão de Barros [a], Raimundo Gerônimo da Silva Júnior [b,c], Eugênio de Sá Coutinho Neto [c], Eid Gonçalves Coelho [c], Carlos Eduardo Coelho de Sá [c]

[a] Health Science Center, State University of Piauí, Teresina, PI, Brazil
[b] Health Science Center, Federal University of Piauí, Teresina, PI, Brazil
[c] Oncology High Complexity Assistance Unit, Dr. Everardo Aragão Macroregional Hospital in Caxias, Caxias, MA, Brazil

Dear editor,

Vulvar sarcomas are rare and represent 1%–3% of cancers in this region.[1] Leiomyosarcoma is the most common type of vulvar sarcoma. In contrast, epithelioid sarcoma (ES) is extremely rare, accounting for around 1% of soft-tissue vulvar sarcomas. Among the ESs of the vulva, proximal variants tend to have a more aggressive clinical course, in comparison to distal variants.[2]

The treatment of choice in proximal-type ES of the vulva is surgical resection, usually with surgical margins greater than 2 cm. Due to the extreme rarity of these malignancies, clear algorithms for adjuvant treatment of these tumors have still not been established. Radiotherapy and/or chemotherapy may be indicated in some cases.[3]

In this study, we present a case of proximal-type epithelioid sarcoma of the vulva, initially misdiagnosed as Bartholin cyst.

A 36-year old woman, with no comorbidities, is referred to our oncology center on August 2021 due to a painful vulvar tumor. The patient reported having two prior vulvar surgeries for Bartholin cysts.

Magnetic resonance imaging (MRI) of the pelvis was performed (Fig. 1). It revealed a large, wide solid lesion, with irregular shape, and intense postcontrast enhancement of the vulva and left perineal region, in intimate contact and no cleavage plane with the ipsilateral adductor muscles, measuring 12 cm. Pelvic MRI findings were consistent with recurrent/residual tumor.

The patient was scheduled for vulvectomy, which was performed the following month (Fig. 2).

According to Fig. 3, histopathology analysis of the surgical specimen showed a solid malignant tumor of epithelioid cells in a multinodular arrangement, with microcysts interspersed and myxoid stroma. Surgical margins were negative. On immunohistochemistry (IHC), there was positivity for epithelial markers (cytokeratin and epithelial membrane antigen – EMA) and CD34 (Cluster of Differentiation *34*) and loss of gene SMARCB1/INI1 expression, confirming the diagnosis of proximal-type ES of the vulva. IHC was negative for S100 protein.

The patient was sent to the Radiology department to receive adjuvant radiotherapy. From January to February 2022, conformational radiation therapy or 3D conformal radiotherapy (3DCRT) was delivered, at a dose of 66 Gy in 33 fractions to the tumor bed and margins. Clinical progress of the patient was good, although she experienced acute radiation-induced toxicity (grade I-II radiodermatitis, grade I enteritis and grade I cystitis). In July of 2022, eleven months after the initial consultation, she is doing well and has periodical clinical follow-up care at the oncology clinic.

According to a study conducted in 2021 by Chung et al.,[4] there were reports of 39 cases of proximal-type ES of the vulva in the literature to date. Due to the rarity of these cases, proximal-type ESs of the vulva are frequently confounded with benign lesions, resulting in misdiagnosis and inadequate patient treatment. Bartholin cyst, benign inflammatory lesions, necrotizing granulomas or other epithelioid cells, malignant rhabdoid tumors, melanomas (including sarcomatoid melanomas) and undifferentiated carcinomas are the main differential diagnoses for

* Corresponding author. Health Science Center, State University of Piau, Teresina, PI, Brazil
E-mail address: rafaelcosta@aluno.uespi.br (R.E.A.R. da Costa).

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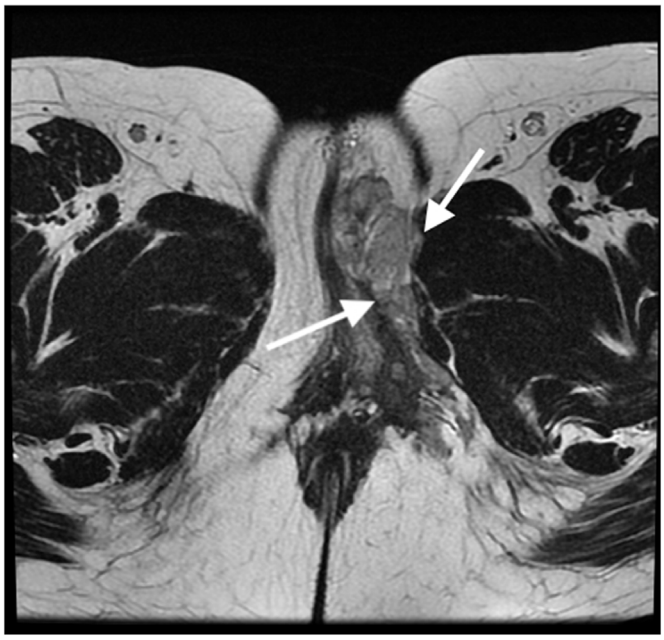

Fig. 1. Magnetic resonance imaging of the pelvis (arrows indicate the tumor).

proximal-type ES of the vulva. The case presented in this study depicts a proximal-type ES of the vulva, initially misdiagnosed and treated inappropriately as a Bartholin cyst.

The mean patient age at diagnosis of proximal-type ES of the vulva is 35 years, similar to what occurs in other sarcomas. The most common symptom in these cases is the presence of a painful and slow-growing mass. Proximal-type ES of the vulva tends to be an indolent tumor. Nevertheless, it has a rapid clinical course and high rates of early metastases, even after surgical resection with negative margins.[3]

Proximal-type ESs of the vulva are usually diagnosed by histopathology. In these cases, IHC should be used to confirm the diagnosis.[5] In IHC, proximal-type ES is usually positive for vimentin, cytokeratin and EMA and negative for S100 and CD31. In some cases, this type of tumor is positive for CD34.[6] The loss of SMARCB1/INI1 gene expression in IHC, as occurred in this case report, is particularly indicative of proximal-type epithelioid sarcoma.[5]

Although radiotherapy is known to be less effective against sarcomas, adjuvant radiotherapy is recommended when there are positive surgical margins and/or advanced tumors. In systemic disease, the recommendation is adjuvant chemotherapy, particularly using doxorubicin and dacarbazine, since the response rate to these agents is most significant in this type of tumor.[3,4]

The case presented in this study is of interest, since it alerts to the importance of early diagnosis of proximal-type ESs of the vulva. In general, despite its extreme rarity, it is a tumor with an aggressive behavior and rapid progression. Furthermore, this case is one of the few cases reported in Latin America, and adds a regional appeal to this issue. In our referral center, a late but correct diagnosis was made. The patient received adequate surgical treatment for an advanced tumor (12 cm) with an indication of adjuvant radiotherapy, due to the initial misdiagnosis. Early surgical tumor resection could have spared the patient from radiotherapy.

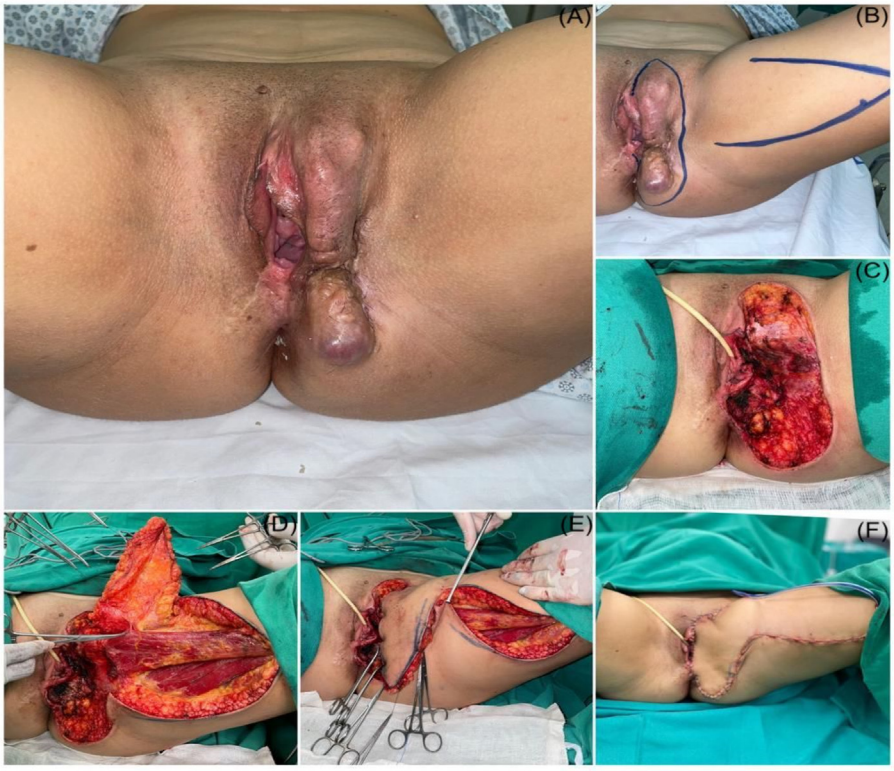

Fig. 2. Surgical resection of proximal-type epithelioid sarcoma of the vulva. (A; B): Preoperative. (C; D; E): Intraoperative. (F): Postoperative.

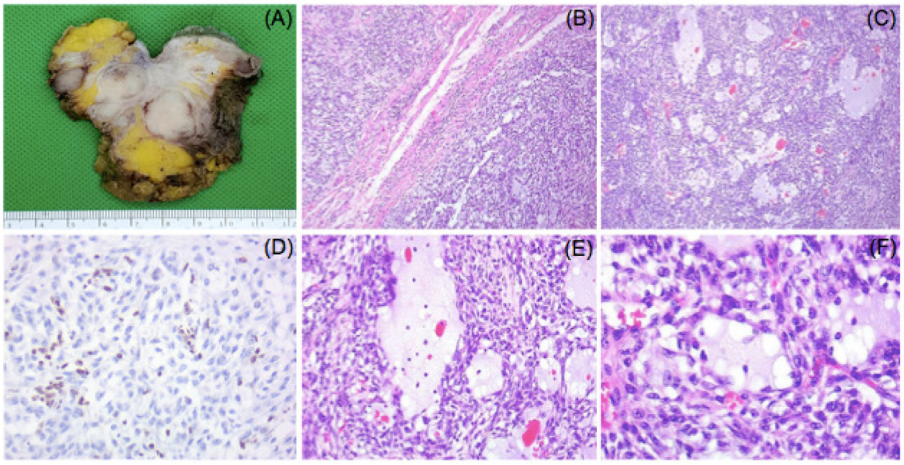

Fig. 3. (A): Surgical specimen. (B; C; E; F): Histopathological HE staining: B:40x; C:100x; E:200x; F:400x). (D): SMARCB1/INI1 gene (magnification: 200x).

Ethical approval and consent

This study was approved by the Institutional Review Board of the State University of Piauí, Teresina (PI), Brazil (CAAE: 54885322.2.0000.5209; reference number: 5.206.809; approval date: 01/20/2022). The patient signed the informed consent form (ICF). All authors consent for publication.

Authors' contribution

All authors have contributed equally.

Declaration of competing interest

We deny any conflicts of interest during the entire performance of this study.

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