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CLINICAL CASE REPORTS

Proximal epithelioid sarcoma of the vulva

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ABSTRACT

Epithelioid sarcoma (ES) is an extremely rare disease that, according to morphological data, can be divided into proximal and distal types. The proximal type of ES (PES) of the vulva arises from the superficial and deep layers of the external genital organs, manifests as single or multiple soft tissue tumor nodules with areas of necrosis and hemorrhage, is characterized by aggressive behavior, and has an unfavorable prognosis due to its high tendency for local recurrence and hematogenous metastasis. Differential diagnosis is performed with various benign and malignant neoplasms, cysts or abscesses of the Bartholin gland, and inguinal and femoral hernias. The final diagnosis and histological type of the tumor are established based on morphological, immunohistochemical, and molecular genetic studies. PES is characterized by solid tumor growth composed of large and pleomorphic epithelioid cells with large vesicular nuclei and distinct eosinophilic nucleoli. Immunohistochemically, loss of SMARCB1 (INI1, BAF47) expression is observed, along with positive expression of EMA, vimentin, and cytokeratins; a positive reaction for CD34 staining is often noted in the absence of expression of other endothelial markers such as CD31 and FLI-1. The clinical presentation and disease characteristics may suggest PES and justify the expansion of the immunohistochemical marker panel. Timely and accurate detection of this tumour plays a key role in improving treatment outcomes, enhancing patients' quality of life, and reducing mortality. However, due to its rarity, PES of the vulva presents objective diagnostic difficulties in both clinical and morphological aspects. Surgical intervention remains the main method of treatment for this pathology, and to date, there are no clearly established recommendations regarding the optimal management strategy for patients with PES of the vulva. This paper presents a clinical case of a 65-year-old patient whose atypical clinical presentation of recurrent vulvar tumor served as the basis for expanding the panel of immunohistochemical markers, which allowed for the diagnosis of PES of the vulva.

Keywords: vulva, sarcoma, proximal epithelioid sarcoma, immunohistochemical study, surgical treatment

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КЛИНИЧЕСКОЕ НАБЛЮДЕНИЕ

Проксимальная эпителиоидная саркома вульвы

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РЕЗЮМЕ

Эпителиоидная саркома (ЭС) является крайне редким заболеванием, в соответствии с морфологическими данными может быть разделена на проксимальный и дистальный тип. Проксимальный тип ЭС (ПЭС) вульвы возникает из поверхностных и глубоких слоев наружных половых органов, проявляется одиночными или множественными опухолевыми узлами мягких тканей с участками некроза и кровоизлияния, отличается агрессивным течением и имеет неблагоприятный прогноз, обусловленный высокой склонностью к развитию местных рецидивов и гематогенных метастазов. Дифференциальный диагноз проводится с различными доброкачественными и злокачественными новообразованиями, кистой или абсцессом бартолиновой железы, паховыми и бедренными грыжами. Окончательный диагноз и гистологический тип опухоли устанавливается на основании морфологического, иммуногистохимического и молекулярно-генетического исследований. Для ПЭС характерен солидный рост опухоли из крупных и плеоморфных эпителиоидных клеток с крупными везикулярными ядрами и легко различимыми эозинофильными ядрышками, при иммуногистохимическом исследовании отмечается потеря экспрессии SMARCB1 (INI1, BAF47), а также позитивная экспрессия EMA, виментина и кератинов, нередко наблюдается положительная реакция на окрашивание CD34 при отсутствии экспрессии других маркеров эндотелия, таких как CD31 и FLI-1. Клиническая картина и особенности течения заболевания позволяют заподозрить ПЭС и расширить панель иммуногистохимических маркеров. Своевременное и точное выявление данной опухоли играет ключевую роль в улучшении результатов лечения, повышении качества жизни пациентов и снижении уровня смертности. Однако из-за своей редкости при ПЭС вульвы имеют место объективные диагностические трудности в клинических и морфологических аспектах. Хирургическое вмешательство остается главным методом лечения этой патологии, и до настоящего времени нет четко установленных рекомендаций по оптимальной тактики лечения больных с ПЭС вульвы. В данной работе представлен клинический случай лечения 65-летней пациентки, у которой нетипичная клиническая картина при рецидиве опухоли вульвы явилась основанием для расширения применяемых для иммуногистохимического исследования маркеров, позволившего диагностировать ПЭС вульвы.

Ключевые слова: вульва, саркома, проксимальная эпителиоидная саркома, иммуногистохимическое исследование, хирургическое лечение

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Соблюдение этических стандартов: в работе соблюдались этические принципы, предъявляемые Хельсинкской декларацией Всемирной медицинской ассоциации (World Medical Association Declaration of Helsinki, 1964, ред. 2013). От пациента получено письменное добровольное согласие на публикацию описания клинического наблюдения.

Финансирование: финансирование данной работы не проводилось.

Конфликт интересов: все авторы заявляют об отсутствии явных и потенциальных конфликтов интересов, связанных с публикацией настоящей статьи.

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BACKGROUND

Vulvar sarcomas are rare malignant mesenchymal tumors characterized by rapid growth, necrotic changes, and a high potential for hematogenous metastasis. According to various authors, sarcomas account for no more than 1–3 % of malignant vulvar neoplasms [1, 2]. Epithelioid sarcoma (ES) is one of the rarest types of vulvar sarcomas and is characterized by aggressive clinical behavior. According to morphological data, ES can be divided into proximal and distal types. This article describes a rare clinical case of proximal epithelioid sarcoma (PES) of the vulva, the clinical course and treatment approaches of which remain insufficiently studied due to the small number of reported cases and the short follow-up period [3, 4].

Description of the clinical case

A clinical case of PES of the vulva in patient S., born in 1959, who underwent treatment at the N. N. Blokhin National Medical Research Center of Oncology. The analysis included data from the medical history, physical examination, laboratory and instrumental studies, morphological and immunohistochemical findings, and long-term treatment outcomes.

The patient presented to the outpatient department of the N. N. Blokhin National Medical Research Center of Oncology in May 2019 with complaints of a mass in the vulvar region. According to the anamnesis, in March 2019 the patient noticed an induration in the area of the left labium majus. At a local medical institution, a fine-needle biopsy was performed, and the cytological picture was interpreted as adenocarcinoma with areas of squamous cell carcinoma. Upon review of the cytological slides at the N. N. Blokhin National Medical Research Center of Oncology, the observed changes were interpreted as melanoma. The patient reported no family history of cancer, menopause since age 52, and two pregnancies in the anamnesis: one full-term delivery and one medical abortion. On physical examination, a painless, mobile, firm-elastic tumor measuring up to 2 cm in diameter was detected, localized in the middle third of the left labium majus.

According to the decision of the oncological council, the patient was hospitalized in the Department of Oncogynecology of the N. N. Blokhin National Medical Research Center of Oncology for surgical treatment. In June 2019, surgical intervention was performed – wide local excision of the vulvar tumor in the form of a left-sided hemivulvectomy and sentinel lymph node biopsy using a radioisotope method.

Macroscopically, the tumor appeared as a mass measuring 2.5 × 2 × 1.5 cm, consisting of confluent nodules of pinkish-gray and yellowish-gray color, soft to firm-elastic in consistency, with whitish fibrous strands. Microscopically, the tumor consisted of solid proliferations of large polymorphic epithelioid cells (Fig. 1), consistent with the structure of melanoma.

Simultaneously with the removal of the vulvar tumor, bilateral excision of the sentinel lymph nodes was performed. To exclude metastatic involvement of the latter, immunohistochemical (IHC) analysis with Melan A, HMB45, and tyrosinase was carried

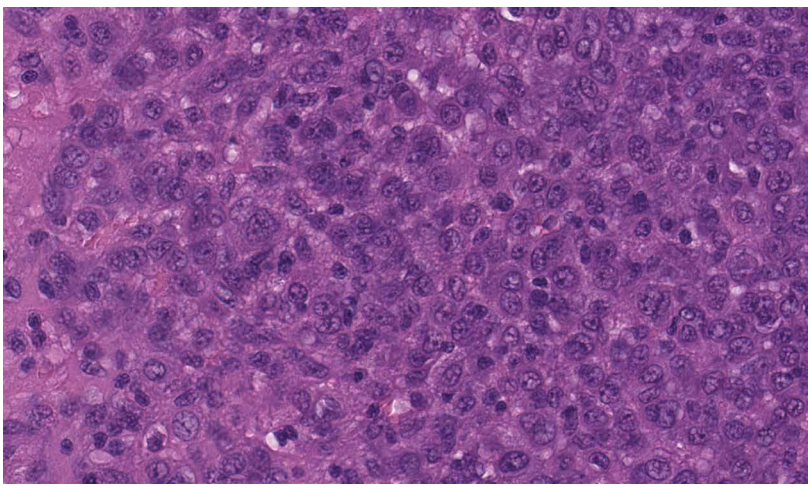

Fig. 1. Pleomorphic epithelioid tumor cells with eosinophilic cytoplasm and enlarged vesicular nuclei with prominent nucleoli (H&E, ×40)

out. No expression of the above-mentioned markers was detected.

Thus, the diagnosis of vulvar melanoma without metastatic involvement of the sentinel lymph nodes was established. Considering the localized nature of the tumor, it was decided to carry out close dynamic follow-up, including quarterly examinations, ultrasound/MRI/CT of the pelvic organs, abdominal cavity, and chest with intravenous contrast enhancement, or whole-body PET-CT.

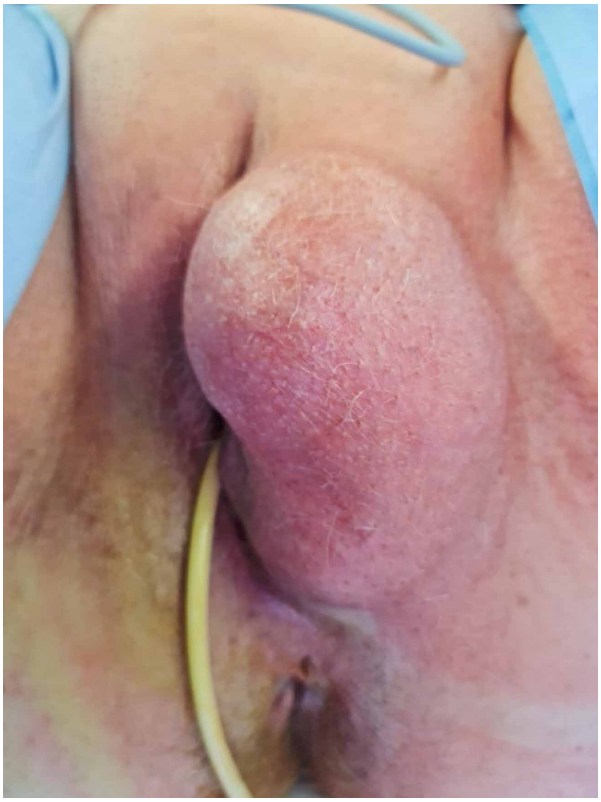

Fig. 2. Recurrent vulvar tumor

In August 2020, the patient noticed the appearance of a nodular mass in the projection of the postoperative scar of the perineal soft tissues. The oncologist at her place of residence referred her for consultation to the N. N. Blokhin National Medical Research Center of Oncology; however, due to the development of coronavirus pneumonia, the patient sought medical attention only in November 2020.

During gynecological examination, deformation of the external genital organs was observed due to the presence of a mass within the soft tissues of the left side of the vulva, of firm consistency, with limited mobility, measuring up to 12 cm in diameter. The skin over the mass was unchanged, and it appeared that the tumor was not associated with the surgical scar (Fig. 2). Examination with vaginal specula was not possible due to compression of the vaginal introitus by the tumor. On palpation, no infiltration of the rectovaginal septum was detected; the rectal mucosa in the projection of the tumor appeared unchanged, and no enlargement or change in the consistency of the inguinal lymph nodes was noted.

According to ultrasound examination of the perineal soft tissues, a lesion with clear, smooth borders and blood flow was visualized within the left labium majus, measuring 11 × 6 cm. MRI of the pelvic organs with contrast enhancement revealed a multinodular lesion of heterogeneous structure

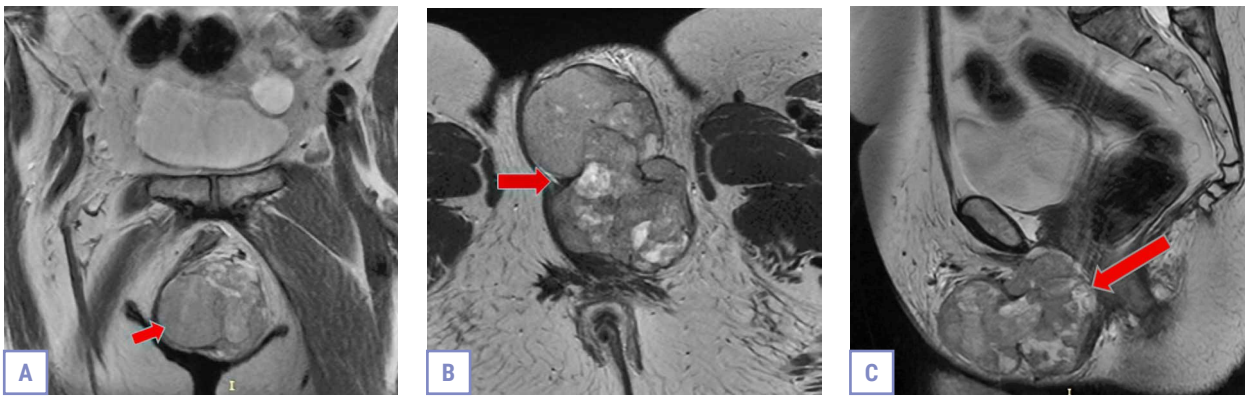

Fig. 3. MRI of the pelvic organs with intravenous contrast enhancement in three projections (A, B, C): a multinodular lesion of heterogeneous structure was detected in the left labium majus (indicated by arrows), measuring up to 55 × 78 × 99 mm, adjacent to the left puborectal muscle and the distal part of the urethra without signs of its invasion.

with well-defined contours, overall dimensions up to 55 × 78 × 99 mm, showing restricted diffusion and active heterogeneous contrast accumulation. The lesion was adjacent to the left puborectal muscle with suspected invasion, to the distal portion of the urethra without signs of invasion, and displaced and deformed the lower third of the vagina without clear evidence of invasion. The pelvic and inguinal lymph nodes were not altered (Fig. 3).

A core biopsy of the vulvar tumor was performed. Histological examination revealed an epithelioid cell tumor, which did not contradict the diagnosis of recurrent epithelioid cell melanoma. However, taking into account the nature of the disease course and the clinical presentation of a mesenchymal tumor without epidermal involvement, additional immunohistochemical (IHC) analysis was performed on the material from the primary tumor using the following markers: S100, HMB45, synaptophysin, chromogranin A, SOX10, CK18, panCK, MelanA, CK7, CD34, BerEP4, Ki67, vimentin, CK20, CD31, desmin, EMA, and FVIII.

Tumor cells showed diffuse and strong expression of vimentin, CD34, and EMA; weak focal expression of CD31, chromogranin A, CK18, panCK, and Melan A. No expression of S100, HMB45, synaptophysin, SOX10, CK7, BerEP4, CK20, FVIII, or desmin was detected in tumor cells. The proliferation index (Ki67) was 24 %. IHC staining with the INI1 marker was not performed at the N. N. Blokhin National Medical Research Center of Oncology laboratory at that time due to technical reasons. However, the co-expression of vimentin, CD34, and EMA, combined with the absence of diagnostically significant expression of markers characteristic of other malignant neoplasms with similar histological structure, allowed the presented morphoimmunophenotype of tumor cells to be interpreted as most consistent with PES.

Comprehensive examination of the patient, including PET-CT, revealed no evidence of regional or distant metastases. Discussion at the oncological multidisciplinary council concluded that, given the local spread of the recurrent tumor, its histological type, and locally aggressive behavior, surgical treatment with a wider margin from the visible tumor boundary was indicated. In December 2020, excision of the recurrent tumor was performed within visually healthy tissues, with a planned margin of at least 2 cm (Fig. 4). The procedure was completed by excision of the labia on the contralateral side to correct the pronounced postoperative deformation of the vulvar region.

The surgical specimen consisted of a fragment of the vulva with adjacent soft tissues measuring

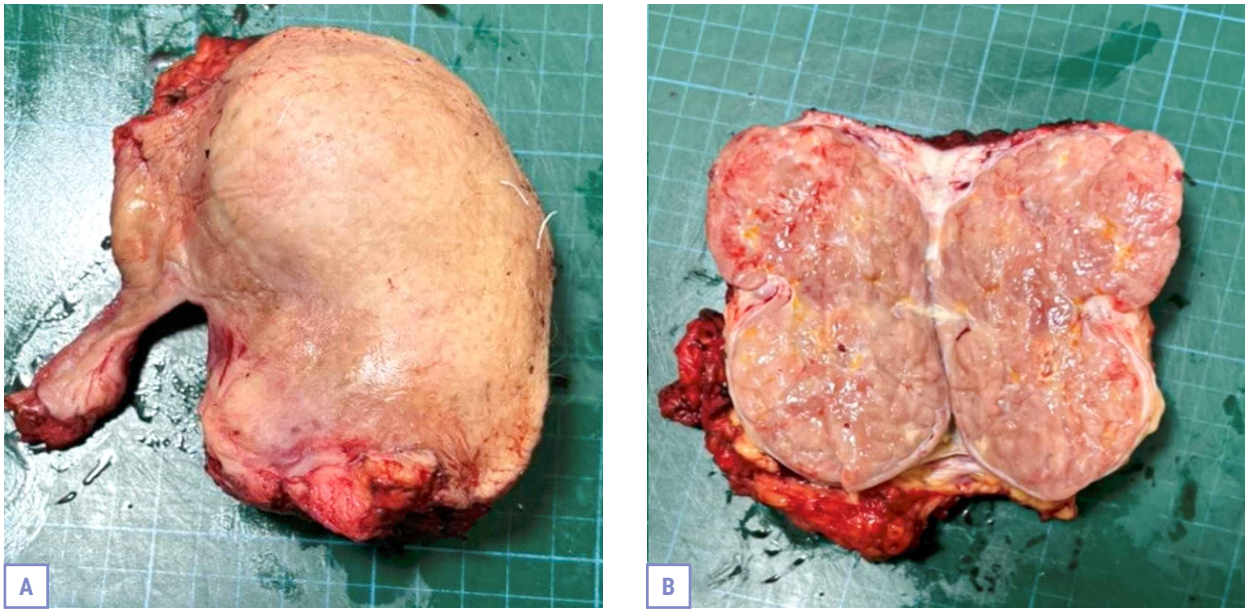

Fig. 4. Gross specimen: A – excised recurrent vulvar tumor; B – tumor on section.

14 × 14.5 × 8.0 cm in total. Within the soft tissues, a subepidermally located tumor nodule measuring 10.5 × 6 × 6.5 cm was identified, with rounded and relatively well-defined borders, composed of confluent lobules of grayish-brown tissue with yellowish streaks and firm-elastic consistency. Microscopically, the tumor was composed of confluent nodules of large epithelioid cells separated by fibrous septa with marked lymphocytic infiltration. Foci of necrosis were observed both in the center and at the periphery of the lobular structures. Mitotic activity reached 20 f. m./10 HPF. The tumor invaded the reticular dermis but showed no invasion of the epidermis and no signs of angiolymphatic or perineural invasion. The morphological pattern was consistent with recurrent PES. No tumor growth was detected at any of the lateral resection margins of the vulva or at the deep margin in the subcutaneous tissue.

Considering the results of the pathological examination of the surgical specimen, the clinical course of the disease, and the absence of evidence of metastatic lesions based on PET-CT findings, the oncological multidisciplinary board decided to continue patient follow-up under the supervision of an oncologist at her place of residence. In December 2023, ultrasound examination revealed a cystic lesion up to 0.5 cm in diameter within the soft tissues in the projection of the left gluteal region. Cytological examination of the aspirate showed no tumor elements. PET-CT with 18F-FDG detected no foci of metabolically active tumor tissue. At present, the patient remains under observation with no signs of recurrence or disease progression.

DISCUSSION

According to the World Health Organization (WHO, 2020) histological classification of tumors, vulvar sarcomas comprise a heterogeneous group of malignant neoplasms differing in clinical course, histological, and immunohistochemical profiles. These include alveolar soft part sarcoma, rhabdomyosarcoma, epithelioid sarcoma, leiomyosarcoma, liposarcoma, dermatofibrosarcoma protuberans, and others [5, 6].

ES is an extremely rare malignant soft tissue tumor with aggressive behavior, which can be divided into proximal and distal types. The proximal type of ES (PES) more commonly occurs in the trunk and pubic area, whereas the distal type typically arises in the upper and lower extremities [7]. PES of the vulva may originate from both superficial and deep tissues, presenting as single or multiple nodules with foci of necrosis and hemorrhage. PES of the vulva is considered a more aggressive form [8]; however, the clinical course of vulvar ES remains poorly understood due to the small number of reported cases and short follow-up periods.

This tumor shares numerous clinical and morphological features with various benign and malignant lesions, including granuloma annulare, melanoma, and epithelioid vascular neoplasms, and may be clinically misdiagnosed as a benign lesion such as a Bartholin gland cyst or abscess, inguinal or femoral hernia, or other benign and malignant soft tissue tumors [9, 10]. In particular, in the present case, the primary tumor was initially interpreted as melanoma, and the clinical presentation did not contradict that diagnosis.

The final diagnosis relies exclusively on pathomorphological examination. PES is characterized by solid growth of large and sometimes pleomorphic epithelioid (carcinoma-like) cells with large vesicular nuclei and distinct eosinophilic nucleoli. Patchy areas of necrosis are often present, but unlike the distal subtype, PES does not form the characteristic pseudogranulomatous pattern. In the distal type, cells show less nuclear atypia, although they may appear more pleomorphic in recurrent or metastatic lesions [11, 12]. Cells with rhabdoid features can occur in both forms but are more frequently seen in the proximal subtype. Immunohistochemical (IHC) analysis serves as a critical tool in the differential diagnosis of ES, with diffuse loss of SMARCB1 (INI1, BAF47) expression and positive co-expression of vimentin, EMA, and cytokeratins being the key diagnostic markers [13]. In a study by Guillou L. et al., more than half of ES cases showed positive staining for CD34 [14], whereas other endothelial markers, such as CD31 and FLI-1, are usually negative [15, 16].

The choice of IHC markers for diagnosis may be of crucial importance. When planning IHC testing, it is essential to consider the clinical presentation

and features of the disease course, which can help expand the marker panel used. In the present clinical case, the atypical appearance of the recurrent lesion, its localization, and the timing of its development after initial treatment prompted the use of an expanded panel of IHC markers, which ultimately allowed for the correct diagnosis.

Cytogenetic studies have revealed chromosomal abnormalities involving the long arm of chromosome 22 in patients with PES, demonstrating inactivation of the tumor suppressor gene SMARCB1/INI1 located at 22q [17, 18].

Chokoeva A. et al. reported that among vulvar sarcomas, ES showed the most aggressive course, while liposarcomas had the most favorable prognosis [19]. According to several studies, unfavorable prognostic factors for ES include primary tumor size greater than 2 cm, deep location, presence of necrosis, high-grade histology, vascular and lymphovascular invasion, early metastasis, and non-radical surgical excision [7, 20, 21]. In a study by Lee N. et al. (2006), an elevated serum CA-125 level was identified as a potentially useful tumor marker for the diagnosis and clinical monitoring of ES [22].

The poor prognosis of patients with vulvar PES is associated with a high tendency for local recurrence, lymph node, and/or distant metastasis. Unlike most other malignant mesenchymal tumors, vulvar ES can also spread via lymphatic and implantation routes to noncontiguous areas of skin, underlying soft tissues, fascia, and bone, which necessitates wider surgical excision [23, 24].

According to a clinical study by Ulutin H. et al., wide excision in the form of vulvectomy prevented local recurrence even without adjuvant therapy. The authors demonstrated that vulvectomy with a wide margin from the visible tumor border provides excellent local control of vulvar ES [25]. However, many authors prefer wide local excision over radical vulvectomy. In a study by Curtin J. et al., only one of seven patients developed local recurrence after surgery alone. Thus, wide excision with a margin of at least 2 cm from the visible tumor border should be performed, as the width of negative resection margins is a major factor influencing local recurrence risk [26, 27]. In the presented clinical case, timely diagnosis of vulvar PES might have allowed for a wider surgical margin during the initial operation, potentially preventing disease recurrence.

Dash B. et al. demonstrated that wide excision remains the preferred treatment for localized PES, while radiotherapy and chemotherapy may be used in unresectable or metastatic forms; however, their role in the adjuvant setting has not been established [28].

Thus, PES represents a pathology associated with objective diagnostic difficulties in both clinical and pathomorphological aspects, which leads to a high rate of diagnostic errors and delays in establishing the correct diagnosis.

CONCLUSION

PES of the vulva is characterized by an aggressive clinical course, necessitating a comprehensive approach to the diagnosis and treatment of this disease. Understanding the morphological, molecular-genetic, and clinical features of vulvar PES contributes to the development of effective therapeutic strategies and improvement of patient prognosis. The presented clinical case clearly demonstrates the importance of timely diagnosis of PES, including immunohistochemical examination, which made it possible to establish the correct diagnosis and plan appropriate treatment.

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