



Oncology

Recurrent vulvar melanoma invading urethra. Clinical case and literature review



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Introduction

Recurrent vulvar melanoma is a rare entity and literature regarding its management is scarce.¹ Mucosal melanomas comprise less than 1% of all melanomas and vulvovaginal melanoma is the second least frequent of them (18%) after those in the urinary tract (3%).² It is the second most common histological type of vulvar cancer, representing 7–10% of all malignant vulvar neoplasms.⁴ Initial symptoms are non-specific and complete excision of the lesion is indicated in cases of suspected diagnosis.¹ Prognosis of patients with these neoplasms is poor and remains unchanged despite the treatment approach.^{1,2} Hemivulvectomy with lymph node dissection is the current procedure of choice, whether combined with adjuvant therapies or not.^{2,3} When affecting the urethra, its behavior is especially aggressive, half of them, presenting with metastasis and with a very short life expectancy.²

We describe a case of recurrent vulvar melanoma invading distal urethra and its management.

Case presentation

A 79-year-old Caucasian woman was followed up by Gynecology and Medical Oncology for a melanoma in the lower third of the left vaginal labia minora that had required hemivulvectomy with selective sentinel node biopsy five years before. Histologic analysis resulted in a superficial melanoma with a maximum Breslow thickness of 1.14 mm. Melanoma recurred two years later and local resection of the left neovulva and new selective sentinel node biopsy was performed. In situ and infiltrating melanoma of the chorion was detected without lymphatic involvement. BRAF mutation was negative, so biochemotherapy was not considered.

Nine months later, she complained of perineal discomfort and dysuria. A whitish nodular, indurated and immobile lesion, painful on palpation and with friable appearance, located in the superior-left paraurethral region was identified (Fig. 1). It was biopsied and pathologic study confirmed that it was a melanoma. Due to its proximity to

the urethral meatus, the Urology Department was consulted. Abdominopelvic CT and PET-CT revealed no locoregional or distant tumor spread. Absence of invasion beyond the urethral meatus was confirmed by urethrocytostcopy.

It was decided to offer the patient a conservative approach with a limited resection, due to the poor prognosis and the doubtful benefit of a more aggressive surgery. We planned to make a distal urethrectomy (Fig. 2). Mucosal defect was covered by an inverted triangular rotational vaginal flap as shown in Fig. 3. The apex of the flap was divided in two to cover the whole urethral meatus. Surgery was uneventful.

Recurrence of vulvar melanoma was confirmed by pathological examination with extensive ulceration, infiltration of the chorion, reaching a maximum thickness of 3.75 mm and extensive areas of melanoma in situ.

Nine months later, there was no evidence of tumor recurrence and the patient had no local or urinary symptoms.

Discussion

Mucosal melanomas constitutes < 1% of all melanomas. The most frequent sites are head and neck (55%), anorectal (24%) and vulvovaginal (18%) and very rarely genitourinary (3%).² When this is the case, the most frequent location is the urethra, especially in its distal part, constituting 4% of urethral cancers.² They are more frequent in white women between the 6th and 7th decade of life.¹ No clear risk factors have been identified, but their location excludes ultraviolet radiation.³

The most frequent symptoms are pruritus, bleeding, palpable urethral mass and micturition difficulty.^{1,3} Differential diagnosis includes chancres, urethral caruncle and other urethral tumors, such as squamous cell carcinoma (50–70% of all urethral cancers), transitional cell carcinoma or adenocarcinoma (10–25%), lymphoma, neuroendocrine carcinoma, sarcoma and paraganglioma.

Physical examination is essential for diagnosis. Body CT, MRI or PET-CT may be useful to evaluate the presence of distant metastasis.¹

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Fig. 1. Amelanotic melanoma in the superior-left paraurethral region.

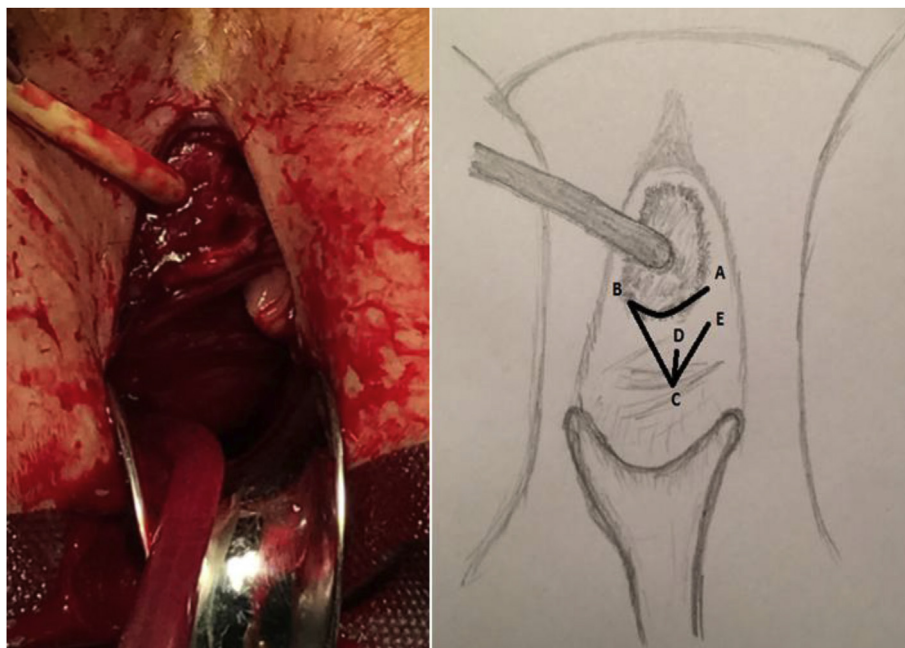


Fig. 2. Urethrectomy.

MRI can differentiate melanoma from other urethral tumors such as carcinoma or leiomyoma because melanin is a paramagnetic molecule, resulting in high T1 intensity images and low in T2.

Diagnosis is confirmed by pathological examination.¹ Approximately 40% of mucosal melanomas are amelanotic, unlike 10% of their cutaneous counterparts, requiring special immunohistochemical techniques such as S-100 protein (highly sensitive) and HMB45 antigen (highly specific).³

Initial management of vulvovaginal melanomas is complete surgical excision and sentinel node lymphadenectomy, which offers the best opportunity for long-term survival.^{2,3} Melanomas with less than 1mm of dermal invasion must be removed with a 1 cm margin, whereas the rest should be excised with a 2–3 cm margin.³ Despite a complete surgical resection, most patients will develop metastatic disease and more extensive resection or radical surgery has not shown to improve overall survival.^{1,2,3} So surgery has to reach a balance between quality of life and aggressiveness to avoid needless morbidity.³

Adjuvant therapy can be added to prevent disease progression.³

However, chemotherapy and radiation therapy are of little value and immunotherapy is only useful if there exists mutations in the BRAF (10%) or the KIT (25%) genes.^{1,2} In that case, imatinib can improve prognosis. Although ipilimumab (anti PD1) has recently proven to significantly prolong survival in cutaneous melanomas, the same has not been demonstrated in mucosal melanomas.

In cases of local recurrence, there is no clear advantage of aggressive surgery versus conservative management. In fact, some authors advocate local control with Imiquimod.⁴ However, the traditional approach has been surgical, with few cases described in the literature. Staiano JJ et al, described a series of 46 patients that required flap reconstruction for gynecological malignancies, 73% of them being recurrences. Complication rate was 53% (wound breakdown in 35% of cases), and 5 year survival was 14%. Only 67% survived one year. The three main flap techniques were rhomboid flap, lotus petal flap and advancement flap.⁵

Due to the poor prognosis of recurrent vulvar melanoma, a conservative approach seems to be a reasonable option. In our case, a

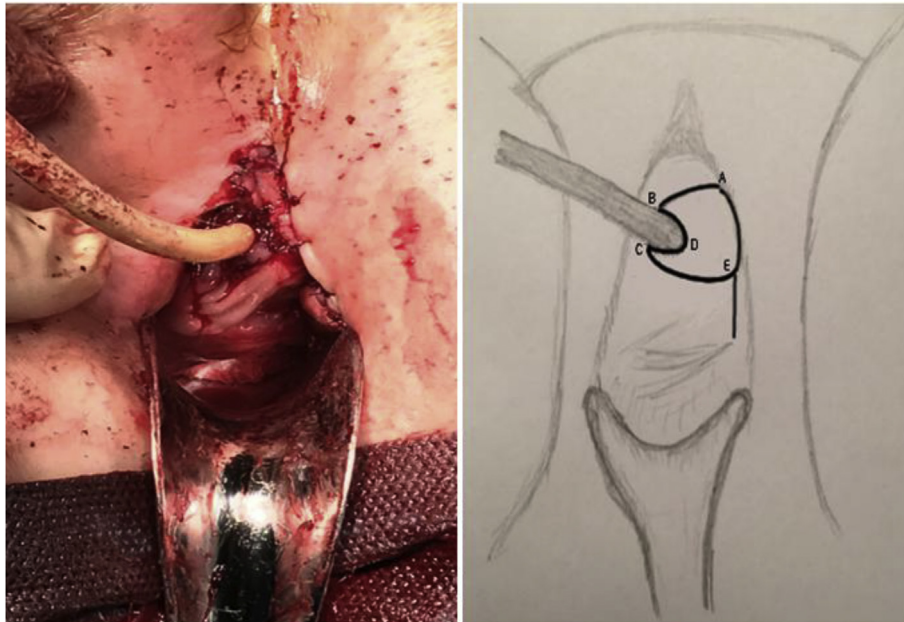


Fig. 3. Advancement vaginal flap.

limited excision and advancement vaginal flap proved to completely control local symptoms with no added morbidity.

Conclusion

Vulvar melanoma is a rare entity with a very bad prognosis. There is no clear way to manage its recurrences, especially when affecting the distal urethra. The benefit of radical surgery should be balanced against the risk of wound complications, always having in mind the short disease-free survival. Vaginal advancement flap is a feasible procedure that can control local symptoms without adding significant morbidity.

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