CASE REPORT

Proximal-type epithelioid sarcoma—unusual presentation: unilateral vulvar mass

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SUMMARY

Epithelioid sarcoma of the vulva is a very rare malignant soft tissue tumour, usually initially asymptomatic, which simulates a variety of benign lesions, leading to diagnosis only at later stages. Prognosis is generally poor. We report a case of a 55-year-old postmenopausal woman, who presented with rapid growth of an asymptomatic mass located in the left labia majora. Biopsy of the lesion revealed features characteristic of proximal-type epithelioid sarcoma. Ganglionic, adrenal, pulmonary and hepatic metastases were found at thoracoabdominopelvic CT scan. During hospitalisation, the patient's clinical condition suffered rapid deterioration and she died on the 16th day of admission.

BACKGROUND

Malignant tumours of the vulvar soft tissue are very uncommon.^{1–2} Of all vulvar malignancies, primary sarcomas occur in only 1–3% of cases.³ Epithelioid sarcoma is a very aggressive soft tissue tumour of mesenchymal origin and was first recognised as a distinct entity in 1970 by Enzinger.^{3–7} Its typical presentation is characterised by a painless nodule or mass.^{1–5} When located in the Bartholin's glands area it can be mistaken for a wide variety of benign lesions, leading to diagnosis at later stages.^{3–6} Early diagnosis is essential to begin appropriate treatment, and thereby improve prognosis and survival.⁵

CASE PRESENTATION

A 55-year-old postmenopausal Caucasian woman, G1P1, was sent to the emergency department of our hospital with symptoms of a rapidly growing asymptomatic mass located at the left labia majora. The referred mass was detected by the patient 2 months earlier but its dimensions were stable until the last 3 weeks, when it began to grow. Anorexia, dyspnoea after medium effort and severe weight loss were also present. No evidence of erythema, bleeding or discharge was noted. The patient had no relevant personal or family history, except for tobacco use since she was 15 years of age.

Gynaecological examination revealed an indurate massive vulvar mass measuring 10×4 cm arising in the left labia majora with a small floating zone (figure 1). The overlying skin showed engorged veins. At palpation, the lesion was fixed and adherent to deep planes. Owing to patient intolerance, speculum observation could not be carried out. Inguinal lymphadenopathy was not detected.

Figure 1 Pelvic inspection: indurate massive vulvar mass (10×4 cm) in the left labia majora.

INVESTIGATIONS

After patient consent, a biopsy of the mass under general anaesthesia was performed. Abundant necrotic and cerebroid material was drained and sent for histological study (figure 2). Subsequently, speculum and bimanual examination unmasked an indurate extensive mass occupying the right upper third of the vaginal wall and posterior vaginal fornix, which was also seen on pelvic ultrasonography (figure 3). Owing to suspicion of Bartholin's gland carcinoma, a CT of the thoracic and abdominopelvic areas was carried out. It revealed a large 8×13×10 cm right paramedian pelvic mass

Figure 2 Vulvar biopsy: abundant necrotic and cerebroid material was drained.

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Figure 3 Pelvic ultrasonography: extensive mass occupying the right upper third vaginal wall and posterior vaginal fornix.

extending from the left labia majora and invading the right ureter, and extending down towards the rectouterine pouch (figures 4 and 5). Regional ganglionic, adrenal, pulmonary and hepatic metastases were also found (figures 6 and 7).

Histological examination showed a set of large cells with epithelioid phenotype and eccentric nuclei (figure 8). The mitotic index of this tumour was 4 per 10 high-power fields. At the immunohistochemical study, tumoural cells were highly positive for vimentin, focally positive for CAM5.2 and negative for S-100 protein, CD34, α -actin and desmin (figures 9 and 10). These findings were compatible with proximal-type epithelioid sarcoma.

DIFFERENTIAL DIAGNOSIS

Clinically, these lesions can be easily mistaken for benign processes, such as Bartholin's cyst, infectious granuloma, lipoma, dermoid cyst, viral warts, fibroma or lipoma. Differential diagnosis based on immunohistochemical findings with other malignant vulvar soft tissue tumours should also be performed, namely, epithelioid peripheral nerve sheath tumour, malignant melanoma, epithelioid angiosarcoma, epithelioid or rhabdoid rhabdomyosarcoma and epithelioid leiomyosarcoma.

TREATMENT

In the present case, despite it being the recommended treatment, surgery was not performed because of the wide dissemination of the disease and the patient's poor general clinical

Figure 4 Pelvic CT (transverse plan): pelvic mass invading the left labia majora (star).

Figure 5 Pelvic CT (transverse plan): pelvic mass (star) invading the bladder (circle).

condition. All possible comfort care was provided to ensure the patient's well-being.

OUTCOME AND FOLLOW-UP

The patient's clinical condition suffered rapid deterioration and she died on the 16th day of admission.

DISCUSSION

Epithelioid sarcoma is a very uncommon soft tissue tumour of mesenchymal origin.^{1 4 5 7} These tumours can be divided into distal or proximal types, occurring, respectively, at upper and

Figure 6 Thoracoabdominopelvic CT (coronal plan): heterogeneous pelvic mass (9.2 cm) invading adjacent tissues, extending to left labia majora, rectouterine pouch and bladder (star); pulmonary nodules suggestive of metastasis (arrow).

Figure 7 Thoracic CT (transverse plan): pulmonary nodules suggestive of metastasis (arrow).

lower extremities or in the trunk, abdomen or pelvis.^{1–4} The proximal type presents a more aggressive behaviour characterised by rapid expanding of the tumour, elevated local recurrence even after surgery with negative margins and propensity for early metastasis.^{4–8}

When located in the vulvar region, it usually affects the labia majora.^{1–3} Until 2012, only about 29 cases had been reported in the English literature.⁵ It usually occurs, similarly to other sarcomas, between the ages of 21 and 57 years, with 35 years being the mean age.^{1–3}

Growth of a painless mass in the labia majora is the most frequent initial symptom.^{1–2} Clinical misdiagnosis with a benign process can result in delay of diagnosis and a worse prognosis.^{1–5}

The tumour can originate from the superficial or deep tissues of the vulva and present as a single or multiple nodules containing focal necrosis and haemorrhage areas.^{1–3} Locoregional lymph nodes can be involved.⁶ Regional metastasis can occur via lymphatic spread or through direct extension, while distant metastases are due to haematogenous dissemination to lung or liver.^{5–6} These tumours characteristically arise out of satellite lesions between the primary tumour and the regional lymph nodes.^{1–3} Macroscopically, the lesions may present as a solid or fleshy white-coloured nodule or mass.^{3–7} If present, necrosis and haemorrhage present a yellow and brown appearance, respectively.⁷ Histological evaluation is necessary for diagnosis.⁹ Microscopically, this tumour can present with different cell

Figure 8 H&E-stained section of the tumour (×400).

Figure 9 Immunohistochemical staining for vimentin (×400).

types, making the correct diagnosis a challenge for the pathologist. The tumour usually presents a nodular arrangement of a mixture of epithelioid and fusiform cells, with central necrosis, reminiscent of the histological characteristics of an infectious granuloma. The cells can be large, have an amphophilic cytoplasm, eccentric nuclei and prominent nucleoli, which may resemble an undifferentiated carcinoma, or may have a 'rhabdoid' differentiation with eosinophilic cytoplasm and a vesicular nuclei, filled by intracytoplasmic inclusions, features difficult to differentiate from a rhabdoid tumour.^{3–7} Abnormal mitotic figures can also be present. A mitotic index in excess of 2 per 10 high-power fields in association with necrosis is adverse microscopic features.³

Immunohistochemistry stainings are useful for differentiating epithelioid sarcomas from other malignant tumours with epithelioid appearance, since different tumours present different combinations of markers. More commonly, epithelioid sarcomas exhibit cytoplasmic immunoreactivity for epithelial markers (cytokeratins and epithelial membrane antigen), mesenchymal markers (vimentin) and CD34.^{5–7} Focally positive reaction to desmin can also be present, but they are negative to S100 and CD31.⁷ Differential diagnosis with malignant melanoma (S100 and HMB positive), epithelioid angiosarcoma (CD31 positive), epithelioid or rhabdoid rhabdomyosarcoma (desmin positive, cytokeratin negative) and epithelioid leiomyosarcoma (desmin and actin positive) should be performed.³

Figure 10 Immunohistochemical staining for CAM 5.2 (×400).

Surgical excision with safety margins remains the recommended treatment.^{1 3-5 7} Prior studies have shown that risk of recurrence is most closely related to inadequate resection of margins (<2 cm) and more often takes place within 6 months after surgery.^{1 2 4 7} Locoregional lymphadenectomy should not be performed by routine, but it is indicated when lymph nodes seem suspicious. Until now, it has not been proven to be beneficial on local or distant recurrence related to lymphadenectomy.^{1-5 7} Adjuvant radiotherapy is advisable in the presence of a high-grade tumour or positive margins.^{2 5} The beneficial effect of adjuvant chemotherapy is not well established, but is recommended in cases of disseminated disease.⁵⁻⁷ Owing to the low incidence of these tumours at this location, there are no evidence-based diagnostic algorithms or published recommendations for treatment.⁷ Decisions are made based on the individual case presentation and pathology evaluation.

Poor prognosis is best correlated to histological high-grade tumours, lymph node involvement, vascular invasion, tumour sizes larger than 2 cm, deep location and presence of necrosis.^{1 4 6} Local recurrence is very common and distant metastases

arise in 60% of cases.^{1 4 6 7} The 5-year overall survival rates for proximal type are worse than for those mentioned for distal type, which vary between 50% and 80%.^{4 7}

Few cases of vulvar epithelioid sarcoma have been reported to date, which is the reason why this case represents another opportunity for sharing orientations in similar cases and improving strategy.

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REFERENCES

- Altundag K, Dikbas O, Oyan B, *et al.* Epithelioid sarcoma of vulva: a case report and review of the literature. *Med Oncol* 2004;21:367–72.
- Ulutin HC, Zelars RC, Frassica D. Soft tissue sarcoma of the vulva: a clinical study. *Int J Gynecol Cancer* 2003;13:528–31.
- Tjalma WA, Hauben EI, Deprez SM, *et al.* Epithelioid sarcoma of the vulva. *Gynecol Oncol* 1999;73:160–4.
- Kim JH, Choy YS, Lee TS. A case of epithelioid sarcoma arising in the vulva. *J Gynecol Oncol* 2008;19:202–4.
- Patrizi L, Corrado G, Saltari M, *et al.* Vulvar proximal-type epithelioid sarcoma: report of a case and review of the literature. *Diagn Pathol* 2013;8:122–7.
- Pai K, Pai S, Sripathi H, *et al.* Epithelioid sarcoma: a diagnostic challenge. *Indian J Dermatol Venereol Leprol* 2006;72:446–8.
- Argenta P, Thomas S, Chura JC. Proximal-type epithelioid sarcoma vs. malignant rhabdoid tumor of the vulva: a case report, review of the literature, and an argument for consolidation. *Gynecol Oncol* 2007;107:130–5.
- Kim HJ, Kim MH, Kwon J, *et al.* proximal-type epithelioid sarcoma of the vulva with INH1 diagnostic utility. *Ann Diagn Pathol* 2012;16:411–15.
- Elkas JC, Berek J. Vulvar cancer: clinical manifestations, diagnosis and pathology. Up-to-date. 2011.

Learning points

- ▶ Primary sarcomas of the vulva account for only 1–3% of all vulvar malignancies.
- ▶ As epithelioid sarcoma presentation can be mistaken for benign lesions or carcinoma, any vulvar lesion with unusual characteristics or insidious evolution in labia majora should be investigated.
- ▶ Early diagnosis is essential to begin appropriate treatment, and thereby improve prognosis and survival.

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