Leiomyosarcoma of the Vulva: a case report

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Summary

Primary sarcoma of the vulva is a rare tumor. Leiomyosarcoma is the most common histologic variant of vulvar sarcoma. The patient, a 55-year-old, gravida 4, para 2 postmenopausal Greek woman, presented with a 5-year history of progressive enlargement of the right labia majora. On vaginal examination there was a 8 x 9 cm raised, ulcerated and irregular mass of the right labia majora. Despite surgery, the patient died six months later because of multiple metastases.

Key words: Vulvar leiomyosarcoma.

Introduction

Primary sarcoma of the vulva is a very rare tumor. The incidence of vulvar sarcoma varies between 1.5 and 5% of all vulvar malignancies. Leiomyosarcoma is the most common histologic type of vulvar sarcoma [1, 2]. It occurs most frequently in the labia majora, followed, in decreasing order, by the Bartholin gland area, clitoris, and labium minus. The mean age is between 35 and 50 years [3].

Case report

A 55-year-old, gravida 4, para 2 postmenopausal Greek woman presented with a 5-year history of progressive enlargement of the right labia majora.

Her past surgical history was significant for anterior colporraphy and total abdominal hysterectomy. Her family history was unremarkable.

On vaginal examination there was a 8 x 9 cm raised, ulcerated and irregular mass of the right labia majora. There were no palpable inguinal lymph nodes, and the rest of pelvic examination was normal.

Preoperative computed tomography (CT) of the chest, abdomen and pelvis, abdominal ultrasound (US), chest X-ray, colonoscopy and uretrocystoscopy were normal.

A wide local excision was performed. Pathology examination of the entire specimen demonstrated a leiomyosarcoma of the vulva, with no free surgical margins and vascular invasion. Tumor cells exhibited high mitotic activity (> 15 mitotic figures per 10 high power fields). Tumor cells were immunoreactive for smooth muscle actin SMA, alfa-smooth muscle actin aSMA (Figure 1), estrogen receptors ER (Figure 2), p53 protein (Figure 3) and ki-67 antigen, but not for CD34 (Figure 4) or cytokeratins.

One month later, the patient underwent radical vulvectomy. Pathology examination of the entire specimen demonstrated free surgical margins and inguinal lymph nodes negative for leiomyosarcoma. The patient declined postoperative radiotherapy.

Four months after surgery palpable inguinal lymph nodes presented (r). CT revealed multiple metastases in the liver and bones.

One month later invasion of the urethra occurred. The patient was in very poor condition. She died seven days later due to multiple metastases.

Discussion

Because of its rarity, our knowledge of this disease has been limited to sporadic case reports and rare reviews, rather than large series. In a large review of the literature, Nielsen *et al.*, found that 36 leiomyosarcomas of the vulva had been reported [4].

The most important pathologic findings for a correct diagnosis are: tumor diameter > 5 cm, an infiltrate margin, a mitotic count of 5 or more per 10 high-power fields and grade 2 or 3 nuclear atypia. If three of these four criteria are fulfilled, leiomyosarcoma is diagnosed, if only two are fulfilled, atypical leiomyosarcoma, and if one or none of the features are present, leiomyoma [4].

The size of the primary tumor and grade are the most important prognostic factors. Favorable prognostic factors have been reported as tumor diameter less than 5 cm, low histologic grade, bladder tumor site, and complete resection [5].

Most authors agree that radical vulvectomy with bilateral lymph node resection is the treatment of choice for sarcoma of the vulva. Whether adjuvant therapy should be offered is still uncertain. Radiation therapy is recommended for cases with high-grade tumor or with margin involvement [6, 7].

Vulvar leiomyosarcoma is relatively more common in the reproductive years. A few cases have been reported in association with pregnancy. This fact provides circumstantial evidence for the involvement of hormonal factors in the pathogenesis of this disease [8].

Conclusion

Because of the small number of patients, we believe that further studies should be done to elucidate the role of hormones in the development and progression of this tumor.

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Fig. 1

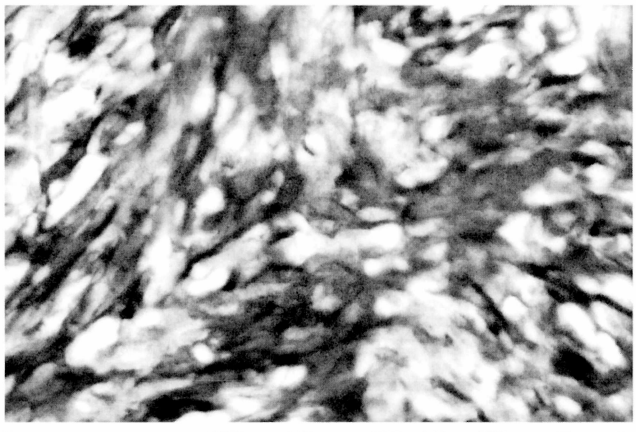

Fig. 2

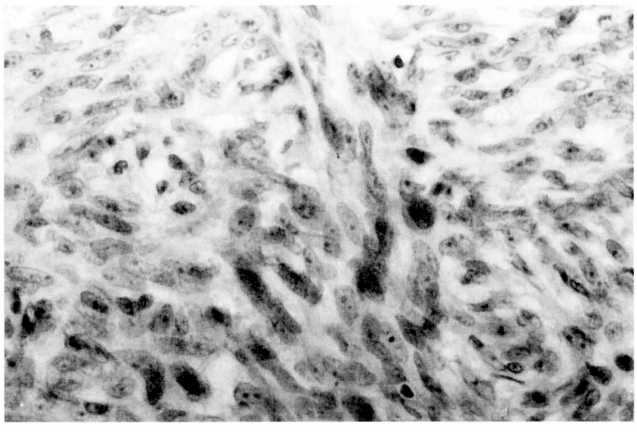

Fig. 3

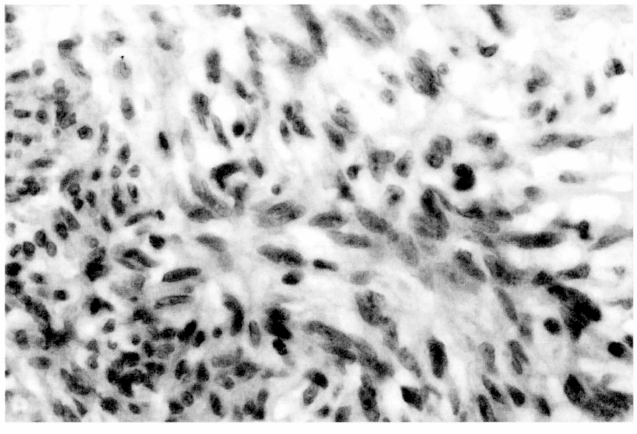

Fig. 4

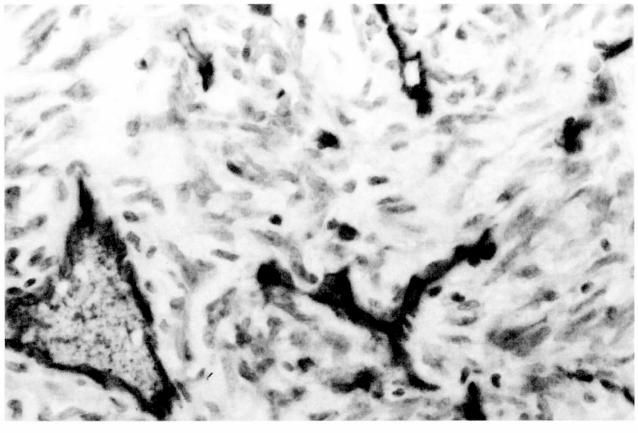

Figure 1. — Tumor cells immunoreactive for aSMA.
Figure 2. — Tumor cells immunoreactive for ER.
Figure 3. — Tumor cells immunoreactive for p53.
Figure 4. — Microvessels, but not tumor cells, immunoreactive for CD34.

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