Leiomyosarcoma of the vulva: a brief communication

L. S. Mensch[1], C. E. Trask[1], G. H. Eltabbakh[2]

Departments of Pathology[1] and Gynecology[2], University of Vermont and Fletch Allen Health Care, Burlington, Vermont (USA)

Key words: Leiomyosarcoma; Sarcoma; Vulva.

Sarcomas represent 1-2% of vulvar malignancies with leiomyosarcoma being the most common [1]. Other soft tissue vulvar sarcomas that have been reported include fibrous histiocytoma, fibrosarcoma, hemangiosarcoma, malignant hemangiopericytoma, epithelioid sarcoma, neurofibrosarcoma, liposarcoma, rhabdomyosarcoma, and malignant schwanoma [1]. Because of its rarity the clinical behavior and management of vulvar leiomyosarcomas are not well established.

A 56-year-old woman presented with an enlarging vulvar mass of one-year duration. The mass was 6 cm in diameter and involved the right labium maius and extended underneath the lower third of the vaginal epithelium. A biopsy revealed a high-grade leiomyosarcoma. Computed tomography of the abdomen, pelvis, and lungs showed no evidence of distant spread. Right radical vulvectomy, partial vaginectomy, and right inguinofemeral lymphadenectomy were performed without complications. The tumor measured 5.5 x 4.2 x 3.3 cm and was whitish-tan, firm, and slightly nodular. Resection margins were negative. The inguinal lymph nodes showed no evidence of metastases.

On hematoxylin and eosin staining, the tumor showed bundles of interweaving spindle cells with a moderate amount of eosinophilic cytoplasm and elongated nuclei with moderate to marked pleomorphism. The average mitotic count was 10-15 per 10 high power fields. The mass had a well demarcated, pushing, noninfiltrating margin (Figure 1). The tumor cells stained positive for alpha-smooth muscle actin (SMA) (Sigma, St. Louis, MO; 1:10,000) and vimentin (Dako, Carpenteria, CA; 1:200), and were negative for HMB45 (Enzo, New York, N.Y., 1:50), S100 protein (Dako, Carpenteria, CA; 1:3,000), and keratin AE1/AE3 (Boehringer Mannheim, Indianapolis; IN; 1:150).

Following surgery the patient received external radiation therapy to a total dose of 6,000 cGy. The patient is alive and without evidence of recurrence 9 months following surgery.

The biologic behavior of soft tissue sarcomas of the vulva is similar to that from other body sites. Prognosis after the appearance of regional or distant recurrence is poor and prevention by excision is the best way to improve prognosis [2].

Tavassoli and Norris reported that recurrences were associated with a diameter greater than 5 cm, infiltrating margins, and greater than 5 mitotic figures per 10 high power fields [3]. In a review of 24 women with vulvar and vaginal sarcomas, Curtin *et al.* reported that grade was the most important predictor of outcome [4]. These authors recommend primary surgical management with adjuvant radiation therapy for high-grade sarcoma and locally recurrent low-grade sarcomas.

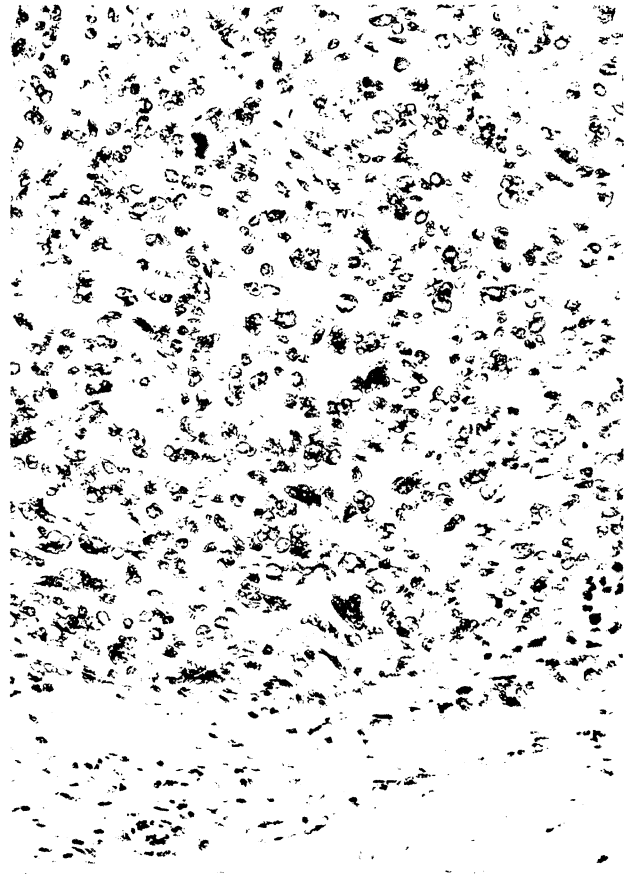

Figure 1. — Tumor (top of photo) demonstrating pleomorphism, a high mitotic rate, and a noninfiltrating, pushing border (Hematoxalin and Eosin, 200x).

References

[1] Fu Y. S., Reagan J. W.: "Benign and malignant epithelial tumor of the vulva". In: Fu Y. S., Reagan J. W., editors, "Pathology of the uterine cervix, vagina, and vulva". Philadelphia, Saunders, 1989, 138.

[2] Aartsen E. J., Albus-Lutter C. E.: "Vulvar sarcoma: clinical implications". *Europ. J. Obstet. Gynecol. Rep. Biol.*, 1994, *56*, 181.

[3] Tavassoli F. A., Norris H. J.: "Smooth muscle tumors of the vulva". *Obstet. & Gynecol.*, 1979, *53*, 213

[4] Curtin J. P., Saigo P., Slucher B., Venkatraman E., Mychalczak B., Hoskins W. J.: "Soft-tissue sarcomas of the vagina and vulva: a clinicopathologic study". *Obstet. Gynecol.*, 1995, *86*, 269.

Address reprint requests to:
LEON S. MENSCH, M. D.
Department of Pathology
Fletcher Allen Health Care
Burlington, VT, U.S.A. 05401

Revised manuscript accepted for publication September 10, 1999