Case Reports

Leiomyosarcoma of the vulva

D. Salehin[1], C. Haugk[1], M. William[2], B. Hemmerlein[2], M. Thill[3], K. Diedrich[3], M. Friedrich[1]

[1]Department of Gynecology and Obstetrics, Helios Hospital Krefeld, Krefeld
[2]Institute of Pathology, Helios Hospital Krefeld, Krefeld
[3]Department of Obstetrics and Gynecology, University Schleswig-Holstein, Campus Luebeck, Luebeck (Germany)

Summary

Malignant tumors of the vulva soft tissue are uncommon. About 1-3% are sarcomas. They can be mistaken as benign lesions, leading to misdiagnosis and mistreatment. A case of a 71-year-old woman with a leiomyosarcoma of the vulva is presented. The surgical excision of the lesion is described and there were no additional malignancies or lesions found. There was no need for adjuvant therapy.

Key words: Vulvar carcinoma; Leiomyosarcoma.

Introduction

Primary sarcomas of the vulva are very rare gynecologic malignancies, accounting for 1-3% of all vulvar malignancies [1-3]. The tumor can be mistaken as a benign lesion as is described in the literature when it occurs in the Bartholin's gland area [4, 5].

Leiomyosarcoma is the most common histologic type of vulvar sarcoma [6], although also reported liposarcomas, neurofibrosarcomas, angiosarcomas and epitheliod sarcomas have been reported [3].

The location of the leiomyosarcoma is in the labia majora, followed by, in decreasing order, the Bartholin gland area, clitoris and labia minora [7].

The age of the patients varies between 31 and 69 years and the size of the tumor varies from 2-10 cm [8].

The case of a patient who had a leiomyosarcoma growth in the left labia majora is reported.

Case Report

A 71-year-old, gravida 3, para 2, postmenopausal German woman presented with a vulvar nodule.

The patient consulted a gynecologist for a preventive medical checkup. She had an unremarkable gynecology history and she had never used any kind of hormonal therapy. The family history was lung cancer of the patient's mother. The gynecology history of the family was unremarkable.

Clinically there was a painless vulvar mass, 2 cm in diameter, which was initially thought to be a myoma. A biopsy had already been done when the woman came to our hospital. Histology showed an infiltrate margin of a tumor composed of interwoven fascicles of spindle cells. The mitotic count was 10 per high-power field and the nuclear atypia was grade 2. A leiomyosarcoma was diagnosed and surgery was planned.

On palpation, there was still a solid mass and a hematoma which was not attached to skin or bone. There were no palpable groin inguinal lymph nodes. The uterus was hardly circumscribable and in ultrasound it appeared enhanced. Computed tomography (CT) of the abdomen followed. The uterus was manifold lobated and the absorption of the barium meal was irregular. The uterus seemed to be necrotic. Malignancy of the uterine tissue could not be ruled out. X-ray of the chest, CT of the liver, coloscopy and urethroscopy were without any pathology.

Preoperatively we planned a wide local excision of the leiomyosarcoma with adequate margins done as a hemivulvectomy of the left side. In addition to this hysterectomy and salpingo-oophorectomy were carried out as well as inguinal lymphonodectomy.

Pathology examination of the entire specimen demonstrated an uterus with multiple myomas encompassing sizes up to 6 cm diameter with no malignant aspects. The ovaries were inconspicuous.

In the preparation of the vulva the intermediate differentiated leiomyosarcoma was seen with free surgical margins. Inguinal and pelvic lymph nodes were negative for leiomyosarcoma. Tumor cells were immunoreactive for smooth muscle actin, desmin and ki-67 antigen.

Because of the direct location to the anus – there were 7 mm between the tumor and anus – radiotherapy was not possible.

Fifteen days after the operation the patient left the hospital in good condition. Close-meshed examinations were suggested.

Discussion

As already mentioned, sarcomas of the vulvar are rare and they appear painless. Symptoms are often only enlarging nodularity and vulvar discomfort. The vast majority of smooth muscle tumors of the female genital tract occur in the uterus, and for this area criteria for distinction between leiomyomas, leiomyosarcomas and smooth muscle tumors of uncertain malignant potential have been formulated.

Clinically leiomyosarcoms can be mistaken as benign processes, such as a Bartholin cyst, infectious granuloma, fibroma, lipoma, or as in our case as myomas [9].

If misdiagnosed, the management is inadequate and the malign tissue expands.

Our knowledge of this disease is limited because of the rarity of these tumors. In 1996 Nielsen *et al*. delineated 36 cases of leiomyosarcomas of the vulva [10]. At least

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Fig. 1

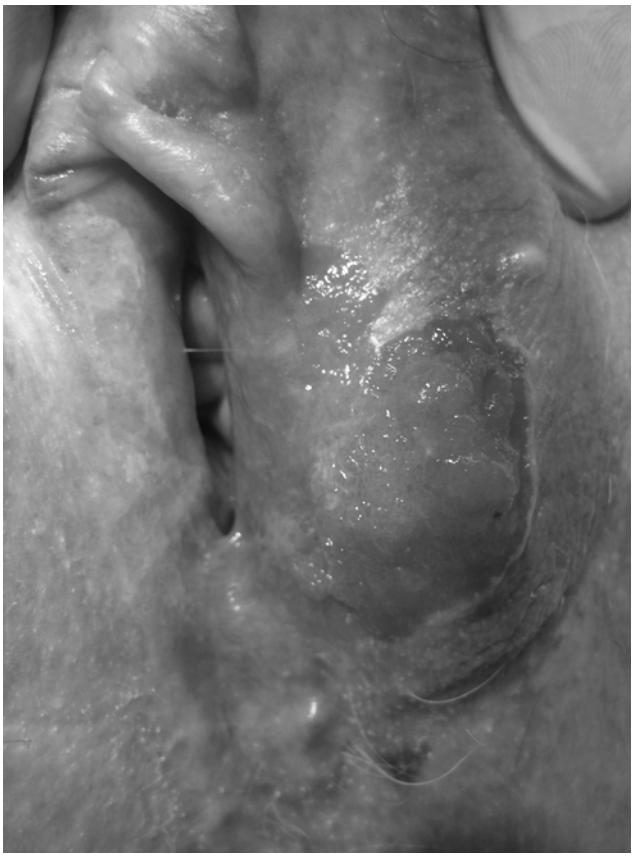

Fig. 2

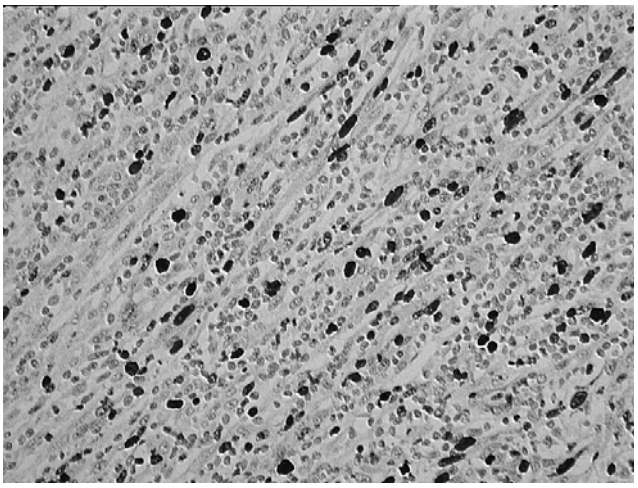

Fig. 3

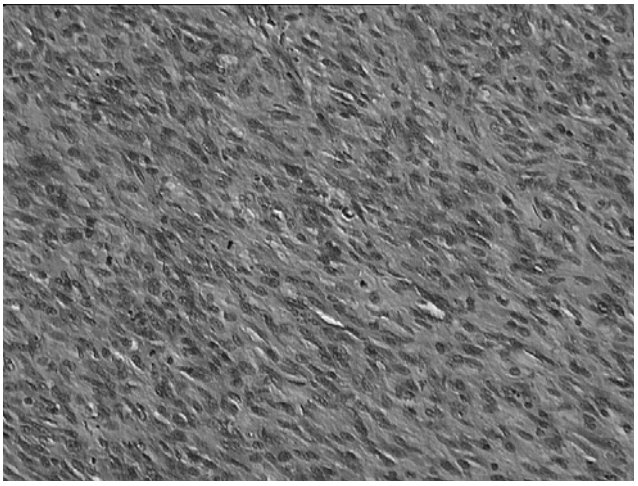

Figure 1. — Vulvar tumor of the left labia majora.

Figures 2, 3. — Tumor composed of interwoven fascicles of spindle cells.

about four times as many leiomyomas have been found in the literature.

Leiomyosarcoma is relatively more common in the reproductive years as seen in a case report during pregnancy of a 36-year-old woman [3].

Diagnostic problems are often caused by these tumors because of the differentiation between benign and malign forms [10, 11]. Nielsen *et al*. reported the most important pathologic findings for a correct diagnosis: 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 two are fulfilled, atypical leiomyoma; and if one or none of the features are present, leiomyoma is diagnosed [10]. Additionally tumor cells of leiomyosarcomas are often immunoreactive for smooth muscle actin and desmin [3]. As prognostic factors the size of the tumor and the grade seem to be the most important [12].

The treatment of choice is radical vulvectomy with lymph node resection as mentioned by several authors and presented in our case [6]. The use of adjuvant therapy is still uncertain. There are cases documented, where radiation is required; these are cases with high-grade tumors, margin involvement or tumor size > 5 cm [13].

In conclusion we would like to mention that this disease is extremely rare and because of the small number of cases further studies should be done. Nevertheless any vulvar lesion with unusual characteristics in the labia majora or Bartholin gland area should be carefully and promptly studied. Based on this, the role of hormones of the tumor should be elucidated because of the higher percentage during reproductive years and especially during pregnancy.

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Address reprint requests to:
D. SALEHIN, M.D.
Department of Gynecology and Obstretics
Helios Hospital Krefeld
Lutherplatz 40
47805 Krefeld (Germany)
e-mail: darius.salehin@helios-kliniken.de