Case report

Primary leiomyosarcoma of the vulva – a rare occurrence

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Vulval tumours are rare, representing merely 4% of all gynaecological neoplasms. 98% of vulvar lesions are benign, and only 2% are malignant. Of all vulvar malignancies, while squamous cell carcinoma is the most common malignancy, leiomyosarcomas of the vulva are extremely rare. Usually these tumours have nonspecific clinical manifestations, often leading to misdiagnoses of Bartholin cysts or abscesses. We describe a case of a 47-year-old woman who presented with a nonspecific painless swelling in the left vulva for 2 months which was diagnosed as leiomyosarcoma of the vulva on biopsy as well as resection.

Key words: vulva, leiomyosarcoma, mesenchymal tumour, genital tract, vagina.

Introduction

Sarcomas of the vulva account for merely 1–2% of all sarcomas of the female genital tract [1]. Primary vulvar leiomyosarcomas (V-LMS) are rare tumours with a poor prognosis [2–7]. They have bimodal age distribution with the first peak occurring at 20–30 years of age and the next in elderly females. These tumours usually have a poor prognosis. As they are located mostly around Bartholin's gland, these tumours are often misinterpreted as a Bartholin cyst or abscess [8, 9]. We present a rare case of V-LMS in a 47-year-old woman.

Case report

A 47-year-old woman presented with left vulval swelling for the last 2 months, gradually increasing in size. It was not associated with any itching, pain, bleeding, dysuria or vaginal discharge. There was no history of significant weight loss or appetite loss. Her menstrual history, obstetric history, past history and family history were normal. On local examination an ill-defined mass was present in the left lower 2/3 of the vulva measuring 8 × 8 cm in diameter, a firm mass with restricted mobility, extending up to the ischiopubic rami but not involving the urethra, clitoris or the underlying bone. Overlying skin was normal. Per speculum examination revealed only a small anterior cervical lip erosion; the vagina was however healthy. PV revealed a normal anteverted uterus along with a firm cervix and bilateral fornixes free and non-tender. The rectal mucosa was free on per rectal examination. Abdominopelvic ultrasound was performed and showed a well-defined round heterogenous mass in the left lower labia, indicating organized content of a Bartholin cyst with vaginal neoplastic aetiology. Vulval biopsy was suggestive of a possibility of LMS. Modified radical vulvectomy with bilateral inguinofemoral lymph nodes' dissection along with reconstruction of the defect with a VY advancement flap was performed.

On gross examination the radical vulvectomy specimen showed a well-circumscribed, soft to firm, grey/white fleshy tumour in the left labia majora and perineum, measuring 6 × 4.2 × 3.6 cm.

Microscopic sections revealed the presence of a spindle cell tumour arranged in bundles and intersecting fascicles with moderate to severe atypia and occasional bizarre forms. There was an occasional focus

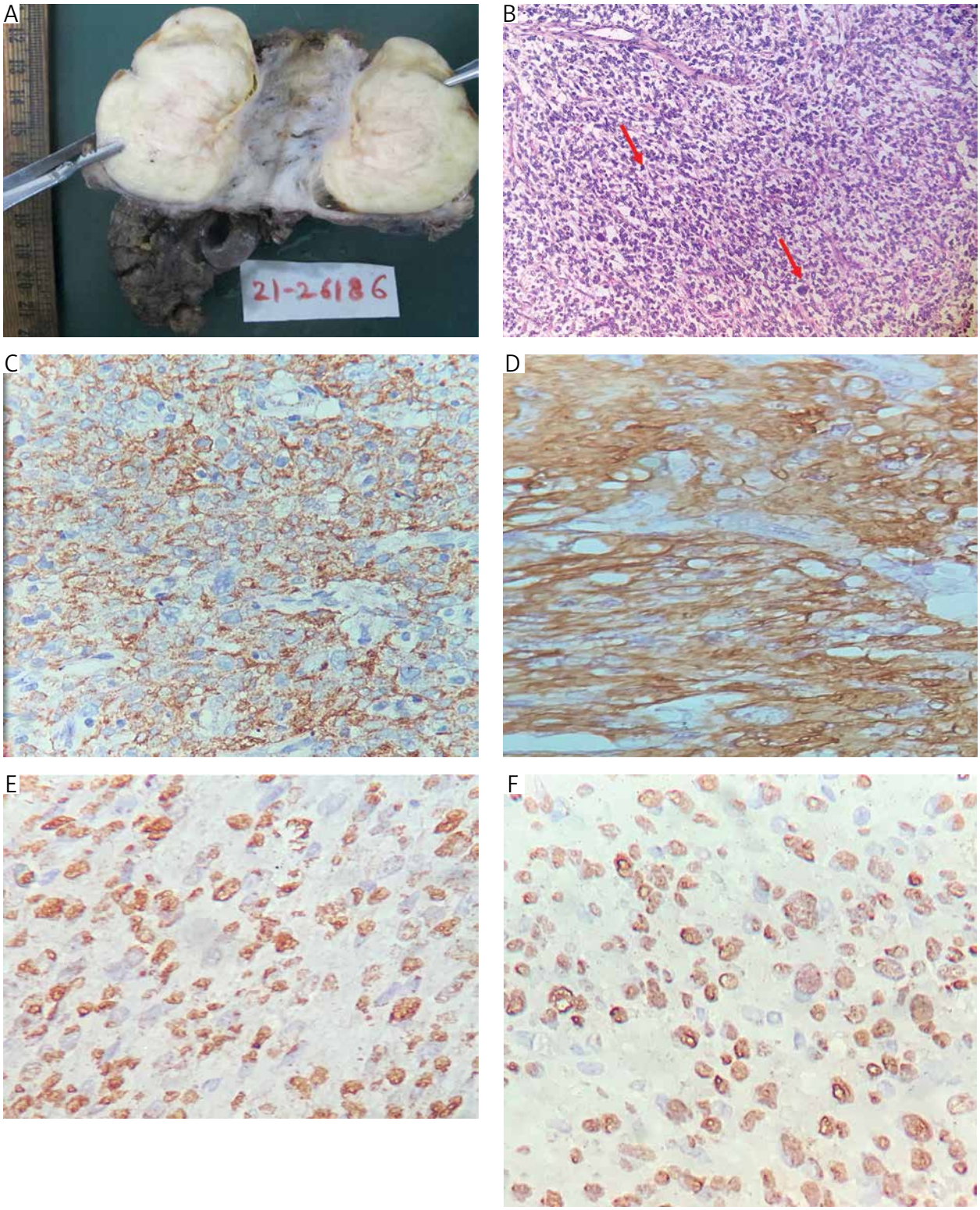

Fig. 1. A) Gross appearance of vulvar leiomyosarcomas (V-LMS); (B) microscopy of V-LMS showing spindle cell tumour with bizarre cells and mitosis (arrows); (C–F) immunohistochemical staining showing immunopositivity with smooth muscle actin (cytoplasmic), desmin (cytoplasmic), oestrogen receptor (nuclear) and progesterone receptor (nuclear) respectively (400×)

of coagulative necrosis with a mitotic count of 6–7/10 HPF. Tumour cells were immunopositive for smooth muscle actin, desmin, oestrogen receptor (ER) and progesterone receptor (PR) and immunonegative for CD117, CD31, CD34 and HMB45. A final diagnosis of leiomyosarcoma vulva was made. The post-operative course of the patient was unremarkable. The patient is disease-free 8 months after surgery.

Discussion

Insidious in onset, V-LMS are rare tumours of the vulva with a poor prognosis [2]. These tumours commonly present as painless, asymptomatic, slow-growing masses, usually in the Bartholin's gland area, and are thought to arise de novo [8]. Depending on the extent of growth they may also present with pain, bleeding or dysuria [1, 5, 7, 8]. However, due to their clinical presentation they are quite frequently misdiagnosed for a benign pathology resulting in treatment delays and worsening prognosis in terms of metastasis [7].

Diagnosis of LMS heavily relies on microscopy, so a prompt biopsy or excision is of utmost importance. The diagnosis of extrauterine LMS according to Nielsen *et al*. requires at least three of the four following criteria:

- diameter ≥ 5 cm,
- infiltrative margins,
- moderate-to-severe (grade 2 or 3) cytologic atypia,
- ≥ 5 mitotic figures per 10 high-power fields (Fig. 1) [1–8].

For patients meeting only one or two of the abovementioned criteria a diagnosis of leiomyoma/atypical leiomyoma is considered. Various factors including mitotic activity, tumour size and grade, pattern of infiltration, presence of necrosis and status of metastasis determine the prognosis in these patients. Immunopositivity for ER/PR indicates the role of hormonal stimulation in the pathogenesis of LMS.

Complete surgical excision of the lesion with adequate negative margins is the recommended mainstay for treatment, though a consensus on the surgical approach is not yet established owing to the rarity of these lesions [9]. Although the data on radiosensitivity or chemosensitivity are still limited, it is advisable to give the same treatment in patients with advanced disease and presence of negative prognostic factors such as incomplete tumour resection, high cellularity, anaplasia, high mitotic activity, severe nuclear atypia and tumour necrosis [1–10].

Conclusions

Vulvar leiomyosarcoma is an uncommon, slow-growing neoplasm with a poor prognosis. A high index of suspicion and prompt biopsy may aid in early detection and better survival for these patients.

The authors declare no conflict of interest.

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