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Primary Vulvar Leiomyosarcoma Presenting as a Cyst-Like Mass in a Middle Aged Woman: A Case Report

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Abstract: ***Background**: Vulvar leiomyosarcoma (VLMS) is the commonest vulvar sarcoma yet represents <1 % of all vulvar malignancies, making evidence-based management challenging. Its ability to masquerade as benign Bartholin-gland cysts or leiomyomas frequently delays definitive treatment. **Case Report**: We report a 46-year-old multiparous woman who presented with a slowly growing, painless swelling of the left vulvo-vaginal vestibule. Baseline laboratory evaluation revealed mild normocytic anaemia with normal biochemical investigations. Pelvic ultrasonography demonstrated a well-circumscribed hypoechoic mass measuring 5.8 × 4.2 × 3.0 cm separated from Bartholin's gland. The lesion was excised with 1 cm macroscopic margins. Grossly the tumour was tan-white, solid and whorled. Histology disclosed a cellular tumor composed of intersecting fascicles of spindle cells with moderate to marked nuclear pleomorphism and brisk mitotic activity (12/10 HPF). Immunohistochemistry was strongly positive for SMA and desmin and negative for S-100, CD34 confirming leiomyosarcoma. Surgical margins were histologically free. Adjuvant radiotherapy was deferred after multidisciplinary review because of clear margins and low to intermediate grade. The patient remains disease-free at 9 months. **Conclusion**: Early complete excision with free surgical margins and histological confirmation is paramount in VLMS because clinical and radiological findings may mimic benign disease. Close surveillance is essential in view of documented late recurrences. This case adds to the limited pool of VLMS reports and highlights contemporary diagnostic and therapeutic considerations.*

Keywords: Vulvar leiomyosarcoma; Bartholin's gland cyst; Vulvar neoplasms

1. Introduction

Leiomyosarcoma (LMS) of the vulva is a rare disease representing 1% of all primary vulvar neoplasms. Still, it is the most common type of vulvar sarcoma [1-3].

The age of patients reported with vulvar leiomyosarcoma at the time of clinical presentation ranged from 14 to 69 years, with mean age of presentation was approximately 30–40 years [4,5,6,7]. The size of vulvar sarcomas varies between 2 to 10-cm [4]. Vulvar sarcomas are often asymptomatic or present with nonspecific clinical features, as an enlarging mass with local discomfort. The tumors arising in the region of the Bartholin's gland are frequently misdiagnosed as benign lesions. As a result, they are commonly mistaken as Bartholin's cyst or Bartholin's gland abscess, which can delay accurate diagnosis and potentially worsen the prognosis [8,9,5]. In such cases, late clinical manifestations may include pain, ulceration, bleeding, and voiding dysfunction [5,8]. Vulvar sarcomas have a high potential for metastasis, and chemotherapy has been reported to induce regression of pulmonary metastases [5,9,7,10].

Vulvar leiomyosarcomas are believed to arise from smooth muscle cells present in structures such as the erectile tissue, vascular walls, round ligaments, dartos muscle, and erector pili muscles, as well as from stem cells located in the region of the Bartholin's gland [11-13]. Leiomyosarcomas most commonly occur in the labia majora, followed by Bartholin's gland region, the periclitoral area, the labia minora [14,15]. Leiomyosarcomas in the region of the Bartholin's gland is extremely rare, with only a few cases reported in the international literature [16,17,18,19,20,9]. The majority of reported cases of vulvar leiomyosarcoma in the international literature involve patients from Western countries, suggesting a possible role of genetic predisposition and lifestyle factors [21]. Chronic inflammation may act as a potential carcinogenetic precursor for these neoplasms. This hypothesis is supported by reports of vulvar leiomyosarcomas coexisting with long-standing lichen sclerosus [22]. Due to the rarity of vulvar leiomyosarcomas, an optimal treatment strategy has not yet been clearly established. Surgical excision remains the primary treatment, which may involve wide local excision or radical hemivulvectomy. The role of ipsilateral lymphadenectomy is unclear, as these tumors predominantly metastasize via hematogenous route rather than through lymphatics. Locally recurrent tumors often require adjuvant radiotherapy, while adjuvant chemotherapy is generally reserved for distant metastases. However, the precise role of adjuvant chemotherapy and radiotherapy remains uncertain, due to aggressive behavior and rapid progression of vulvar leiomyosarcomas [17,18,19].

We report a rare case of vulvar leiomyosarcoma arising in the Bartholin's gland region in a 46-year-old female, highlighting its histopathological and immunohistochemical characteristics. Additionally, the current literature is reviewed regarding clinical presentation, diagnostic approaches, biological behaviour, prognosis and treatment strategies. Vulvar sarcomas in the Bartholin's gland area are frequently misdiagnosed as benign lesions, which can lead to delayed diagnosis and management.

2. Case Presentation

A 46-year-old multiparous woman with no significant past medical history presented with a five-month history of a gradually enlarging, painless swelling in the left vulvo-vaginal vestibule, unaccompanied by pruritus, discharge, constitutional "B" symptoms or menstrual

irregularity. Physical examination revealed a firm, mobile submucosal mass measuring approximately 6 cm; with no ulceration or palpable inguinal lymphadenopathy. Baseline investigations demonstrated normal complete blood count and an essentially unremarkable liver function test, renal function test, coagulation profiles and both fasting and post-prandial blood glucose levels. VDRL and HIV serology were non-reactive. High-frequency trans-perineal ultrasonography showed a well-circumscribed, predominantly solid, hypoechoic lesion measuring 5.8 × 4.2 × 3.0 cm, containing focal cystic areas and minimal internal vascularity, lying separate from Bartholin's gland and with no sonographic evidence of pelvic nodal disease or uterine or adnexal abnormality. The mass was excised via a longitudinal vestibular incision under spinal anaesthesia with a macroscopic 1 cm margin. The encapsulated specimen (5.5 × 4.0 × 3.0 cm) had a tan-white, solid, whorled cut surface (Figure: 1). Histology revealed an infiltrative malignant mesenchymal tumour, composed of long intersecting fascicles of spindle cells. Cells show moderate to marked nuclear pleomorphism having plumped vesicular to hyperchromatic nuclei and pale eosinophilic cytoplasm. Mitotic count was 12 /10 HPF. Atypical mitotic figures present (Figure: 2). On Immunohistochemistry spindle cells were diffusely positive for smooth-muscle actin(SMA) and desmin, focally positive for h-caldesmon, negative for S-100, CD34 and cytokeratins (Figure: 3). Finally we established a Fédération Nationale des Centres de Lutte Contre le Cancer (FNCLCC) grade 2 vulvar leiomyosarcoma. All circumferential margins were free by approximately 4.0 mm. Post-operative recovery was uneventful, and a multidisciplinary tumor board recommended close surveillance without adjuvant therapy because of intermediate grade and negative margins. The patient remains asymptomatic and disease-free nine months after surgery on a follow up advice of three-monthly clinical examinations and six-monthly imaging.

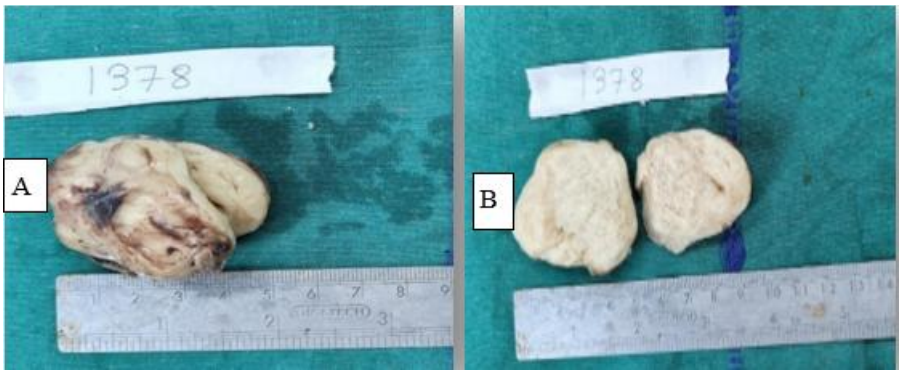

Figure 1: (A) Gross specimen image showing a single, yellowish white, globular bosselated solid tissue measuring (5.5x4.0x3.0) cm. (B) Cut surface image shows solid, firm, whorled appearance

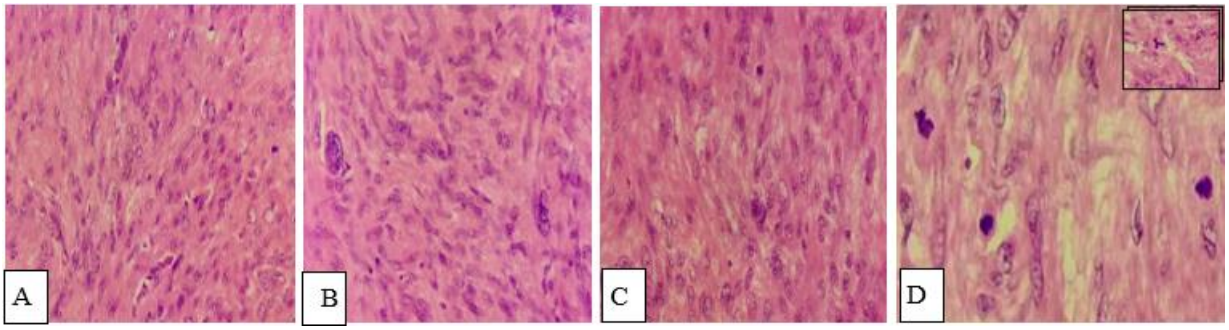

Figure 2: (A) cellular neoplasm composed of intersecting spindle cells with hyperchromatic nuclei (H and E, 400x); (B, C) Moderate to marked nuclear atypia with hyperchromatic nuclei and coarse chromatin (H and E, 400x); (D) Numerous mitotic figures with atypical mitotic figures (inset) (H and E, 400x).

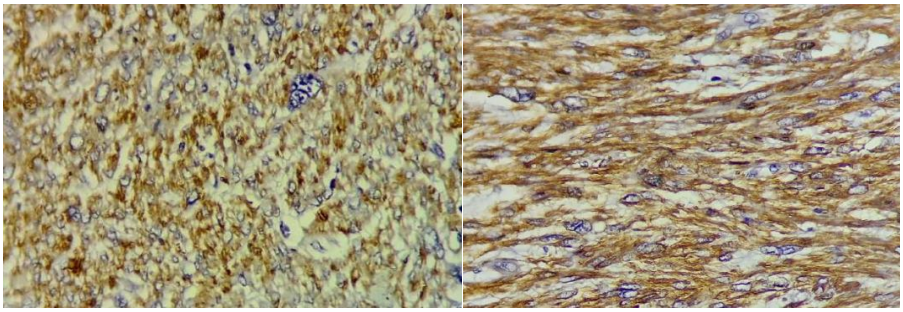

Figure 3: (A) Spindle cells show diffuse cytoplasmic positivity for Desmin (400x) and (B) Strong cytoplasmic positivity for SMA (400x).

3. Discussion

The incidence of vulvar sarcomas varies between 1.5% and 5% of all vulvar neoplasms, with LMS being the most frequent histological type [17, 23]. Other vulvar sarcomas include embryonal and alveolar rhabdomyosarcoma, epithelioid sarcoma, alveolar soft part sarcoma, and other rare sarcomas [9, 24]. Isolated cases and limited series of LMS cases have been described in literature and the largest series (25 cases) was reported by Aartsen and Albus- Lutter [16].

These tumors typically follow an insidious course, most often presenting as a gradually enlarging, painless solid mass located in the labia majora or labia minora. In some cases, patients may also experience pain, pruritus, or erythema. The definitive diagnosis of leiomyosarcoma is usually established through histopathological examination of the excised tissue, as clinically the lesion is frequently misdiagnosed as a Bartholin's gland cyst or as a leiomyoma when the mass has a firm consistency. The definitive diagnosis of leiomyosarcoma is usually established through histopathological examination of the excised tissue, as clinically the lesion is often mistaken for a Bartholin's gland cyst or a leiomyoma when the mass is firm. Soft-tissue smooth muscle tumors that demonstrate both cytologic atypia and significant mitotic activity are generally classified as leiomyosarcomas, reflecting their potential for metastatic behaviour [25]. Diagnostic criteria to differentiate between leiomyomas and leiomyosarcomas of the vulva have been suggested. Tumors exhibiting three or more of the following characteristics should be regarded as leiomyosarcoma: (1) > 5.0 cm in diameter, (2) infiltrative margins, (3) > 5 mitoses/10 HPF, and (4) moderate to severe cytological atypia. Tumors exhibiting only one of these characteristics should be diagnosed as leiomyoma, and those exhibiting two of these characteristics should be regarded as benign but atypical leiomyoma [26]. Leiomyosarcoma must be considered in the differential diagnosis of other soft tissue tumors. The histological grade is the most important prognostic factor in soft tissue sarcomas, as it is closely associated with the likelihood of metastasis and overall patient survival. Among the available grading methods, the FNCLCC grading system is the most widely documented and well-validated system [27].The presence of smooth muscle actin, h-caldesmon, and desmin positivity confirmed the diagnosis of vulvar leiomyosarcoma in the Bartholin's gland area. Multiple case reports have described patients presenting with slowly enlarging, painless vulvar masses that were initially presumed to be Bartholin's cysts. However, subsequent biopsy revealed these lesions to be malignant, highlighting the potential for delayed diagnosis and curative treatment due to initial clinical misinterpretation. [15, 28, 6, 11]. Due to the rarity of these tumors, there are no established evidence-based diagnostic algorithms or standardized treatment guidelines. However, previous reports have suggested several management approaches, including wide local excision, hemivulvectomy, ipsilateral inguinal lymphadenectomy, chemotherapy, and radiotherapy [29, 20]. Management decisions are typically made on an individual basis, taking into account the clinical presentation and pathological findings. Leiomyosarcomas are generally treated with complete surgical excision, with the aim of achieving histologically negative margins. In our patient, the mass was excised based on the initial clinical diagnosis of a chronic Bartholin's cyst. Histopathologic examination and immunohistochemistry have confirmed the case as vulvar leiomyosarcoma. Extensive local excision with free margins of 4 mm without radiation therapy was performed on this patient. Nine months later, the patient is well with no evidence of recurrence.

4. Conclusion

In conclusion, any vulvar lesion with unusual characteristics in the Bartholin's gland region should be thoroughly evaluated, as vulvar leiomyosarcoma (VLMS), though rare, should remain as a major differential diagnosis for solid vulvar lesions in middle-aged women. For VLMS, wide local excision with clear surgical margins and histopathological examination with immunohistochemistry allow accurate diagnosis and achieve durable local control in most of the cases. Ensuring adequate initial surgery is crucial to prevent local recurrence and distant metastasis.

In cases of recurrence, repeat extensive surgical resection combined with ipsilateral lymphadenectomy and radiotherapy is recommended, while chemotherapy is reserved for distant metastases. In the absence of adverse prognostic factors, no need for radical surgery or adjuvant therapy. Given the potential for late local recurrence, close long-term follow-up is essential to enable timely detection and management of recurrent disease.

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