

CASE REPORT/OLGU SUNUMU

A rare mass on the localization of Bartholin gland: Leiomyosarcoma

Bartholin bezi lokalizasyonunda nadir görülen bir kitle: Leiomyosarkom

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ABSTRACT

Vulvar leiomyosarcoma is a rare malignant tumor of the vulva originating from smooth muscle, yet it is the most common type of vulvar sarcoma. It often presents as a benign lesion, which can lead to misdiagnosis and delays in appropriate treatment. We present a case that was initially suspected to be an anal abscess or Bartholin gland abscess. Imaging revealed a solid vulvar mass with fibroid-like characteristics rather than a cystic structure. Following wide local excision, histopathological examination and immunohistochemical staining confirmed the diagnosis of vulvar leiomyosarcoma. Current literature emphasizes the lack of a standardized treatment algorithm due to the rarity of this tumor. This case highlights a personalized treatment approach enabled by interdisciplinary collaboration.

Keywords: leiomyosarcoma, Vulvar sarcoma, Vulvar leiomyosarcoma, Bartholin cyst

ÖZET

Vulvar leiomyosarkom, vulvanın düz kas dokusundan kaynaklanan nadir bir malign tümördür; ancak vulvar sarkomlar arasında en sık görülen tiptir. Genellikle iyi huylu bir lezyon izlenimi verdiğinden, yanlış tanıya ve tedavide gecikmelere yol açabilir. Bu yazıda, başlangıçta anal apse veya Bartholin apsesi olarak değerlendirilen bir olgu sunulmaktadır. Görüntüleme yöntemleriyle yapılan incelemede, kistik yapıdan ziyade miyomu andıran solid özellikte bir vulvar kitle saptanmıştır. Kitlenin geniş lokal eksizyonunun ardından gerçekleştirilen immünohistokimyasal boyama ve patolojik inceleme sonucunda vulvar leiomyosarkom tanısı konmuştur. Güncel literatürde, vulvar leiomyosarkomların oldukça nadir görülmesi nedeniyle standart bir tedavi algoritması bulunmadığı vurgulanmaktadır. Bu olguda, disiplinler arası iş birliğiyle hasta bazlı kişiselleştirilmiş bir tedavi yaklaşımı benimsenmiş ve vaka, mevcut literatürdeki yönetim stratejileriyle birlikte değerlendirilmiştir.

Anahtar Kelimeler: Leiomyosarkom, Vulvar sarkom, Vulvar leiomyosarkom, Bartolin kisti

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INTRODUCTION

In gynecological practice, soft tissue and visceral sarcomas are rare tumors, accounting for an estimated 4% of all gynecological malignancies.¹ They most frequently arise in the uterus (83%), followed by the ovaries (8%), vulva and vagina (5%), and other gynecological organs (2%)²

Leiomyosarcomas are classified as soft tissue sarcomas (STS) of smooth muscle origin, arising from the smooth muscle component throughout the body. They originate from mesenchymal tissue and typically occur in intra-abdominal organs, the retroperitoneum, and the walls of blood vessels.² They are not considered a distinct gynecological malignancy. Gynecologists are generally more familiar with uterine leiomyosarcomas than those occurring in the vulva.

Vulvar leiomyosarcomas (VLMS) are extremely rare, representing only 1% to 3% of all malignant vulvar tumors.³ However, VLMS is the most common histological type among primary vulvar sarcomas.¹ It exhibits a bimodal age distribution in women, with the first peak occurring between 20 and 30 years of age and the second in women over 50.⁴ The youngest reported case was 14 years old, and the oldest was 72.⁴ VLMS most frequently occurs in the labia majora, followed by the Bartholin gland, clitoris, and labium minus, respectively.³

Clinically, VLMS typically presents as a painless mass near the Bartholin gland, often leading to an initial misdiagnosis as a Bartholin cyst or abscess.⁵ Due to its rarity, there is currently no standardized guideline for the management of VLMS. Consequently, clinical decision-making relies primarily on case reports published in the literature. Herein, we report a case of VLMS and review the current literature regarding preoperative evaluation, diagnosis, treatment

options, and follow-up.

CASE REPORT

A 44-year-old virgin woman presented with a growing mass on the left side of her vulva. She reported progressive swelling accompanied by pain while sitting, which had developed over the past six days. She had no history of systemic illness or prior surgery. The patient initially visited the general surgery outpatient clinic with a suspected anal abscess. However, following a rectal and anal examination, the general surgeon ruled out an anal abscess and referred her to a gynecologist with a presumptive diagnosis of a Bartholin abscess.

Gynecological examination revealed a well-defined, palpable mass measuring 7×6×7 cm on the left side of the vulva, located at the 5 o'clock position on the left labium majus. No palpable lymphadenopathy was detected. Transabdominal ultrasound showed a normal uterus and ovaries. On translabial ultrasound, the mass appeared more consistent with a myoma-like structure rather than a cystic lesion. Subsequently, contrast-enhanced pelvic MRI was performed. It revealed a solid mass with significant contrast enhancement, extending superiorly toward the bladder base, laterally to the left vaginal wall, and inferiorly toward the rectum. No lymphadenopathy was identified in the pelvic or inguinal regions.

Symptomatic treatment with a cephalosporin-group antibiotic and an anti-inflammatory agent was administered for one week. With this treatment, the edema and tenderness subsided. However, the patient continued to experience discomfort while sitting, so local excision was planned.

The surgery was performed under spinal anesthesia. A vertical skin incision was made

over the mass, followed by sharp and blunt dissection. Intraoperatively, the vulvar skin and subepithelial layer appeared unremarkable. A solid, well-defined mass was located in the subcutaneous tissue, in the region of the left Bartholin's gland (**Figure 1**). Rectal examination confirmed that the rectum and anal canal were free of involvement. Vaginal examination could not be performed due to the patient's virgin status.

The mass was completely excised from the surrounding tissues in a manner similar to a myomectomy. It did not resemble an abscess. As frozen section analysis was not available, the final diagnosis was deferred until the permanent pathology results were obtained. The skin was then closed subcutaneously using 3-0 Monocryl absorbable sutures.

The patient's postoperative period was uneventful, and she was discharged 24 hours later. Final pathology revealed a vaginal leiomyosarcoma (VLMS) measuring 6×7×7 cm with positive surgical margins. Immunohistochemical staining showed focal positivity for Caldesmon, Actin, CD99, BCL-2, Desmin, Pancytokeratin (PSK), CD10, and p63. Stains for HMWCK, CK-7, CK-20, HMB-45, CD34, S-100, and CD68 were negative. The Ki-67 proliferation index was 70%. Due to the malignant pathology findings, a whole-body PET-CT scan was performed. The imaging showed no evidence of metastatic disease; however, a residual mass with high uptake measuring 10×7 mm was detected at the corner edge of the left vaginal canal. The case was discussed at an interdisciplinary tumor board.

The patient was informed that there is no established standard guideline for the management of VLMS. She declined both

radiotherapy (RT) and hemi-vulvectomy. Ultimately, re-excision of the surgical margin was decided. The residual disease was removed with a minimum of 5 mm tumor-free resection margins. The tumor cells were positive for SMA, Desmin, and Caldesmon. The Ki-67 proliferation index remained at 70%, and the mitotic index was 16 mitoses per 10 high-power fields (HPF). The resection margin was confirmed to be tumor-free. Final tumor staging was pT2 pNX L0 V0 R0 G2. The patient was discharged four days later. Adjuvant external beam radiation therapy (EBRT) was planned. She underwent the recommended regimen of 25 fractions of 2 Gy (total 50 Gy), with a boost of 2 Gy × 5 fractions (total 10 Gy) to the former tumor bed, bringing the total to 60 Gy. The patient was followed up with pelvic MRI and thoracic CT every three months. No recurrence was observed during the 12-month follow-up period.

Written informed consent was obtained from the patient for publication of this case report and related images.



Figure 1: External view of leiomyosarcoma during excision

DISCUSSION

The risk of any vulvar lesion being malignant is very low, as 98% of vulvar lesions are benign and only 2% are malignant. The most common vulvar malignancy is squamous cell carcinoma, while vulvar sarcomas are

extremely rare.² Approximately 80 subtypes of soft tissue sarcomas are recognized.⁶ Well-defined subtypes of vulvar sarcomas include malignant fibrous histiocytomas, dermatofibrosarcomas, leiomyosarcomas, angiosarcomas, angiomyxomas, liposarcomas, chondrosarcomas, rhabdomyosarcomas, and epithelioid sarcomas.⁷ The rarity and heterogeneity of these tumors often lead to delays in diagnosis and treatment. Mortality rates have reportedly increased in Europe due to limited progress in prevention, diagnosis, and therapy.⁸

Vulvar leiomyosarcomas (VLMS) present with nonspecific clinical features and are frequently misdiagnosed as Bartholin cysts or abscesses.⁵ VLMS typically causes discomfort due to pressure exerted by the growing mass, although it may be painless in the early stages. As the tumor enlarges, pain may develop. Since VLMS originates in the subcutaneous tissue of the vulva, a solitary fibrous nodule is often palpable upon examination.⁵

The non-cystic, solid nature of the mass and the absence of inflammatory signs should prompt clinicians to consider a potential vulvar malignancy rather than a benign Bartholin cyst in the differential diagnosis. In this case, perineal ultrasonography initially raised suspicion of a tumor. This is a non-invasive, simple, and cost-effective method that should be utilized in initial evaluations. In the second stage, contrast-enhanced pelvic MRI findings further supported the possibility of a malignant vulvar tumor. The relationship between MRI features and tumor grade is well established: irregular, multilobulated tumor shapes, intertumoral heterogeneity, and peritumoral contrast enhancement are commonly associated with high-grade soft tissue sarcomas.⁹

In the preoperative work-up, percutaneous biopsy is often not effective for diagnosing vulvar leiomyosarcoma (VLMS), as it may yield benign cytological findings. This can result in delayed or inappropriate treatment and a poorer prognosis. For an accurate diagnosis of VLMS, total excision of the nodule followed by pathological examination and immunohistochemical (IHC) staining is essential.¹⁰ Pancytokeratin, desmin, smooth muscle actin, and caldesmon are frequently positive markers in VLMS.¹¹ Percutaneous biopsy may be considered when rapid initiation of treatment is necessary, particularly in cases involving metastatic disease where neoadjuvant therapy is planned.¹¹ Given the rarity of vulvar leiomyosarcoma, there is no universally accepted treatment protocol.¹² Classification and staging of VLMS are based on the guidelines for soft tissue sarcomas. Therapeutic recommendations follow the management protocols outlined by the European Society for Medical Oncology (ESMO) and the National Comprehensive Cancer Network (NCCN).¹²

The primary treatment for vulvar leiomyosarcoma (VLMS) involves wide local excision of the tumor with negative resection margins. In extensive or invasive cases, radical hemivulvectomy may be required.¹² Routine inguinal or pelvic lymph node dissection is not recommended.¹² There is no universally defined minimal surgical margin for VLMS. Since these tumors are often attached to adjacent organs such as the bladder, vagina, rectum, or anus—as in our case—“only” clean margins may be considered acceptable in certain situations.¹³

If pathology reveals high-grade VLMS, radiation therapy may be indicated. However, it is generally not necessary for low-grade VLMS.¹² In cases with positive surgical margins, re-

excision aiming for a 2 cm clear margin or hemivulvectomy can be performed.¹² If negative margins are successfully achieved, three additional cycles of adjuvant chemotherapy and radiation therapy may be administered.¹² The administration of chemo therapy is considered to prevent the risk of recurrence or metastasis.¹²

Tumor size and grade are important prognostic factors.^{6,8,11} Neoadjuvant chemotherapy is supported on the basis that it can enable radical surgery by reducing tumor size. One published case involved a 72-year-old woman with a 12.5 cm VLMS who received three cycles of neoadjuvant chemotherapy, resulting in a partial response. This was followed by radical vulvectomy with negative surgical margins. Three additional cycles of adjuvant chemotherapy were administered, and no recurrence was observed during the follow-up period.¹⁴ If the tumor appears infiltrative or metastases are detected, neoadjuvant chemotherapy combined with radiotherapy

may be considered.¹⁴ Due to the invasive nature of these tumors, the risk of recurrence remains high—even with negative surgical margins—ranging from 65% to 77%.⁶ Additionally, local recurrence in low-grade tumors may also warrant adjuvant radiotherapy.¹²

Management of VLMS during pregnancy requires balancing the risk of tumor progression with the risk of premature delivery.¹⁵ Tumor size has been reported to increase rapidly during pregnancy. If feasible, complete resection of the tumor is recommended at 34 weeks of gestation under spinal anesthesia. Following pathological confirmation of vulvar leiomyosarcoma, induction of labor should be planned. Vaginal delivery may be permitted if there are no obstetric contraindications. Postpartum imaging with contrast-enhanced abdominopelvic MRI and PET-CT is used to exclude local or distant metastases.¹⁵

The different clinical scenarios for the management of VLMS are summarized in Table 1 with relevant references.

Table 1: Clinical Scenarios and Management of Vulvar Leiomyosarcoma (VLMS)

Scenario/Case	Diagnostic Approach	Treatment Strategy	Additional Notes	Reference
Initial presentation (non-specific symptoms)	Physical exam, perineal ultrasonography	Proceed with further imaging and diagnostic evaluation	Symptoms may mimic Bartholin cyst; solid, non-cystic mass without inflammation should raise suspicion	[5]
Suspicion of malignancy	Contrast-enhanced pelvic MRI	Guides surgical planning; MRI features like multilobulation, heterogeneity indicate high-grade tumors	MRI is helpful in assessing tumor grade and extent	[9]
Preoperative work-up	Total excision with pathological and IHC analysis	Preferred over percutaneous biopsy	IHC markers (pancytokeratin, desmin, SMA, caldesmon) are frequently positive in VLMS	[10,11]

Localized or extensive VLMS	Pathology and imaging to assess tumor extent	Wide local excision with negative margins; radical hemivulvectomy for extensive cases	Clean margins acceptable in some cases involving adjacent organs	[12,13]
		Re-excision with 2 cm margin or hemivulvectomy; adjuvant chemotherapy and radiotherapy (3 cycles) if negative margins achieved		
Cases with positive or high-risk margins	Pathological confirmation	Adjuvant chemotherapy and radiotherapy (3 cycles) if negative margins achieved	Adjuvant therapy aims to reduce recurrence/metastasis risk; lymph node dissection not	[12]
Large, infiltrative, or metastatic tumors	Clinical staging, biopsy if needed	Neoadjuvant chemotherapy ± radiotherapy, followed by surgery	Helps reduce tumor burden before radical surgery	[6,11,14]
Case example (72 y/o, 12.5 cm tumor)	Biopsy, followed by imaging	3 cycles neoadjuvant chemo → radical vulvectomy → 3 cycles adjuvant chemo	No recurrence observed in follow-up	[14]
Pregnancy with VLMS	Clinical evaluation + imaging	Surgical excision at 34 weeks under spinal anesthesia, followed by induction of labor	Vaginal delivery possible if no obstetric contraindications; postpartum PET-CT & MRI to rule out metastases	[15]

CONCLUSION

This case presents the successful management of vulvar leiomyosarcoma through local re-excision and radiotherapy following a positive surgical margin after wide local excision. It highlights the diagnostic challenges and emphasizes that treatment can be individualized based on the radiologic and pathologic characteristics of the tumor.

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