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VULVAR LEIOMYOSARCOMA MASQUERADING AS A BARTHOLIN GLAND CYST: A RARE DIAGNOSTIC CHALLENGE

Pathology

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ABSTRACT

Background Appearances can be deceptive in surgical pathology. Vulvar leiomyosarcoma is an exceptionally rare malignant smooth muscle tumor that frequently masquerades as benign vulvar lesions, particularly Bartholin gland cysts. This clinical disguise often delays diagnosis until histopathological examination reveals its true malignant nature. We report a rare case of vulvar leiomyosarcoma that highlights the diagnostic challenges posed by this uncommon entity and emphasizes the indispensable role of histopathology and immunohistochemistry in achieving an accurate diagnosis. **Case Presentation** We reported the case of a 46 year old woman presenting with a painful vulvar swelling of two months duration. Clinical examination and ultrasonography suggested a Bartholin gland cyst. Following surgical excision, histopathological examination revealed an infiltrative spindle cell neoplasm composed of intersecting fascicles of pleomorphic cells with brisk mitotic activity (12/10 HPF). Immunohistochemistry demonstrated diffuse positivity for Smooth Muscle Actin (SMA) and Desmin, confirming the diagnosis of vulvar leiomyosarcoma. **Conclusion** This case highlights how a seemingly innocuous vulvar swelling can conceal an aggressive mesenchymal malignancy. It underscores the indispensable role of meticulous histopathological examination and immunohistochemistry in establishing the diagnosis and guiding appropriate management. Reporting such uncommon presentations not only expands the existing literate but also reinforces the need to consider rare malignant entities in the differential diagnosis of persistent vulvar masses, ultimately facilitating earlier recognition and improving patient outcomes.

KEYWORDS

Vulvar Leiomyosarcoma, Bartholin Gland Cyst, Spindle Cell Tumor, Immunohistochemistry

Introduction

"*Not every vulvar swelling is a Bartholin gland cyst*". Hidden behind the appearance of a common benign lesion may lie an aggressive soft tissue sarcoma. Vulvar leiomyosarcoma is an exceptionally rare malignant smooth muscle tumour, accounting for approximately 1% of all primary vulvar malignancies, yet it represents the most common subtype of vulvar sarcoma.(1-3) Because it usually presents as a slow-growing painless vulvar mass, it is frequently mistaken for benign conditions such as Bartholin gland cyst, lipoma, or leiomyoma, often delaying definitive diagnosis.(2-4)

The diagnosis ultimately depends on meticulous histopathological examination supported by immunohistochemistry, as clinical and radiological findings are often nonspecific.(2,4,5) Given its propensity for local recurrence and distant metastasis, early recognition and complete surgical excision are crucial for improving patient outcomes.(3,5) We reported a rare case of vulvar leiomyosarcoma that clinically masqueraded as a Bartholin gland cyst, highlighting the diagnostic challenges and the indispensable role of pathology in unveiling this deceptive malignancy.

Case Presentation

A 46 year old second gravida female presented with complaints of pain and progressive swelling in the vulvar region for two months. There was no significant past medical or surgical history.

Ultrasonography demonstrated a solid heterogenous lesion measuring 56x40 mm in the lower vulva near the Bartholin gland region. Based on clinical and radiological findings , a provisional diagnosis of Bartholin gland cyst was made

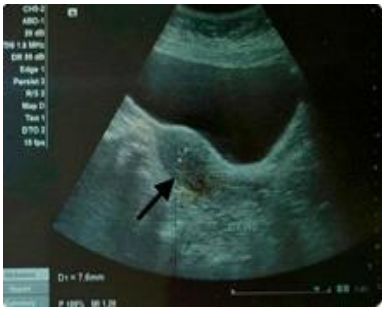


Figure 1: Radiography shows a heterogenous mass

The patient underwent surgical excision under spinal anaesthesia, and the specimen was submitted for histopathological examination

Pathological findings

Gross Examination

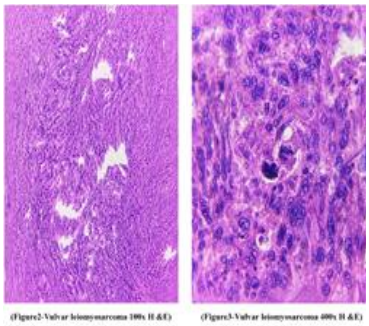
The specimen consisted of a single skin-cvered yellowish- white bosselated solid tissue mass measuring 5.8x4.2x1.3 cm. Cut section revealed a solid grey- white whorled appearance without obvious cystic degeneration

Figure: Gross picture of Vulvar Leiomyosarcoma

Microscopic examination

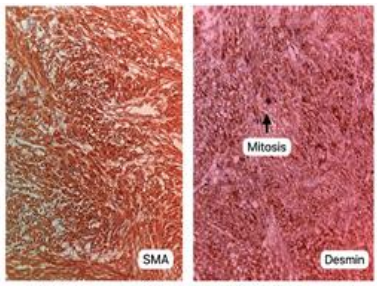
Histological sections demonstrated an infiltrative spindle cell neoplasm composed of intersecting fascicles of elongated spindle-shaped cells with eosinophilic cytoplasm and cigar shaped nuclei. The tumor exhibited marked nuclear peomorphism and hyperchromasia.

Mitotic activity was brisk with approximately 12 mitoses per 10 high power fields, including atypical mitotic figures. Tumor infiltration into the surgical margins was identified. No definite tumor necrosis or haemorrhage was observed.



Immunohistochemistry

The tumor cells showed diffuse cytoplasmic positivity for Smooth Muscle actin (SMA) and Desmin, confirming smooth muscle differentiation and supporting the diagnosis of vulvar leiomyosarcoma.



(Figure4-Vulvar leiomyosarcoma-SMA) IHC (Figure5-Vulvar leiomyosarcoma-Desmin) IHC

DISCUSSION

"Vulvar leiomyosarcoma" is often described as a great masquerader because its clinical presentation rarely reflects its biological aggressiveness. A slowly enlarging vulvar mass in a middle aged woman is far more likely to be labelled as a Bartholin gland cyst, lipoma, or leiomyoma than a malignant spindle cell neoplasm. Consequently , the diagnosis is frequently established only after histopathological examination, as occurred in our patient(2,6)

Our case illustrates an important diagnostic pitfall. Despite clinical examination and ultrasonography favouring a benign Bartholin gland cyst, the excised specimen revealed classical histomorphological features of leiomyosarcoma, including diffuse cytological atypia, brisk mitotic activity (12/10 HPF), infiltrative growth, and positive surgical margins. The diffuse expression of SMA and Desmin further confirmed smooth muscle differentiation while excluding several histological mimics.(3,4)

The rarity of vulvar leiomyosarcoma often results in a low index of clinical suspicion. The diagnostic criteria proposed by Nielsen et al.- tumor size> 5cm, infiltrative margins, moderate-to-severe atypia, and >5 mitoses per 10HPF- remain invaluable in distinguishing leiomyosarcoma from atypical leiomyoma and other spindle cell tumors.(7) Our case fulfilled multiple criteria, firmly supporting the diagnosis.

Beyond establishing the diagnosis, this case underscores a broader principle in surgical pathology: common clinical diagnoses do not always predict common pathological outcomes. Every excised vulvar lesion deserves meticulous histopathological evaluation, regardless of its seemingly benign clinical appearance. Missing this diagnosis may lead to inadequate surgical management , increasing the risk of local recurrence and distant metastasis.(2,5)

Although radical surgical excision with negative margins remains the cornerstone of treatment , vulvar leiomyosarcoma is associated with unpredictable behavior and prolonged risk of recurrence. Therefore, careful postoperative surveillance is essential. By documenting this unusual presentation, we hope to increase awareness among clinicians and pathologists that even a lesion clinically indistinguishable from a Bartholin gland cyst may conceal an uncommon but potentially aggressive malignancy.

CONCLUSION

Rare tumors often test clinical judgement by imitating common benign conditions, and vulvar leiomyosarcoma is a classical example of this diagnostic paradox. Our case highlights that careful pathological evaluation should never be overlooked, even when clinical and radiological findings appear reassuring. Maintaining a high index of suspicion, supported by histomorphology and immunohistochemistry, is crucial for timely diagnosis and appropriate management. Every rare diagnosis reported today broadens the collective clinical experience and improves recognition of similar cases in the future.

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