Vulval leiomyoma: a rare clinical entity

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DESCRIPTION

Vulvar leiomyoma is a rare and benign lesion that can affect women of any age group. It is often misdiagnosed as a Bartholin cyst preoperatively, which make them clinically masquerading. Vulvar leiomyomas represent about 3.8% of all benign soft tissue tumours.[1 2] They usually arise from deeper connective tissues of labia majora (dermatofibroma).[1] Vulvar leiomyoma grows slowly and usually presents as a mass in the perineal region with pain and discomfort as the primary symptoms.

We present the case of a middle-aged woman with no significant past medical history. She attained menarche at the age of 13 years and had regular cycles. The patient presented to the clinic with a 1-year history of a mass per vagina. The patient developed pain since 1 month over the perineal region. On local examination of the vulva, there was a firm pedunculated mass measuring 10×4 cm located at the anterior surface of the left side of the labia majora (figure 1). Routine blood investigations were within normal limits. Pelvic ultrasonography revealed a vulvar growth with features suggestive of neoplastic changes. On Ultrasonography(USG) pelvis, no other fibroid was to be found. Uterus was normal in size and texture, and bilateral ovaries were normal. Fine needle aspiration cytology (FNAC) revealed a mass of spindle cells. The patient was planned for a surgical excision of the vulvar mass. Postoperatively, gross specimen evaluation revealed well-circumscribed mass measuring approximately 10.5×4 cm, with areas of necrosis suggesting secondary degenerative changes (figure 2). Histopathological examination revealed a benign changes composed of smooth muscle spindles arranged in whorled and parallel bundles, admixed with inflammatory cells (figure 3). Final pathological analysis concluded it to be a vulvar leiomyoma with secondary degeneration. Long-term follow-up at 1, 3 and 6 months showed no evidence of recurrenc.

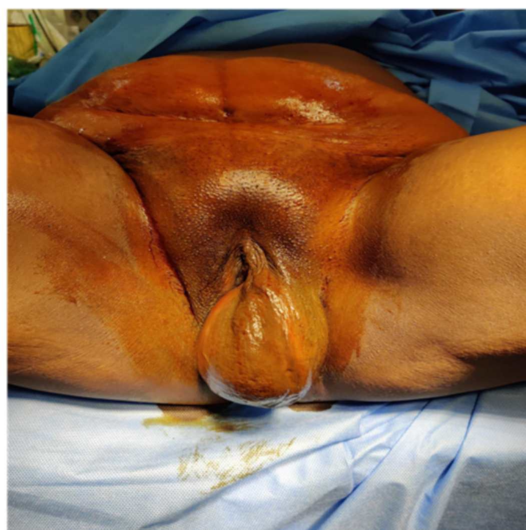

Figure 1 Local clinical examination image of mass per vagina.

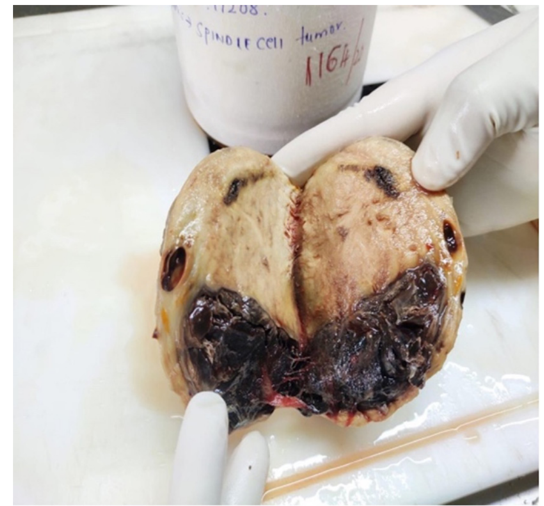

Figure 2 Gross postoperative specimen showing areas of necrosis.

Review of literature suggests less than 200 case reports that have been presented so far.[1 3] Leiomyomas are benign tumours having multiple sources of histological origin including smooth muscle cells, spindle cells and epithelioid cancer cells of eosinophilic cytoplasm. Often, a Bartholin's cyst is a common alternative misdiagnosis to vulval leiomyoma as they both share similar presenting symptoms, such as painless mass per vagina, erythema, discomfort and localised swelling of the area.[2 4 5] Clinical features such as everted labia minora and cystic consistency of the swelling suggest the possibility of a Bartholin cyst; however, finding inverted labia minora and firm consistency of the swelling support the diagnosis of vulvar leiomyoma. Transperineal ultrasonography and pelvic MRI can be used for diagnostic purposes.

Benign versus malignant leiomyoma lesions is another diagnostic challenge. In the article published by Neielsen and colleagues, they proposed a criteria to resolve the aforementioned challenge based on a four-point perspective.[6] This included more than 5 cm in widest dimension, infiltrative margins, more than five mitotic figures per 10 hpf, and moderate to severe cytologic atypia. If three or all features were found, then the neoplasm is considered to be a sarcoma. Benign but atypical leiomyomas fulfil only two characteristics, and benign leiomyomas are the

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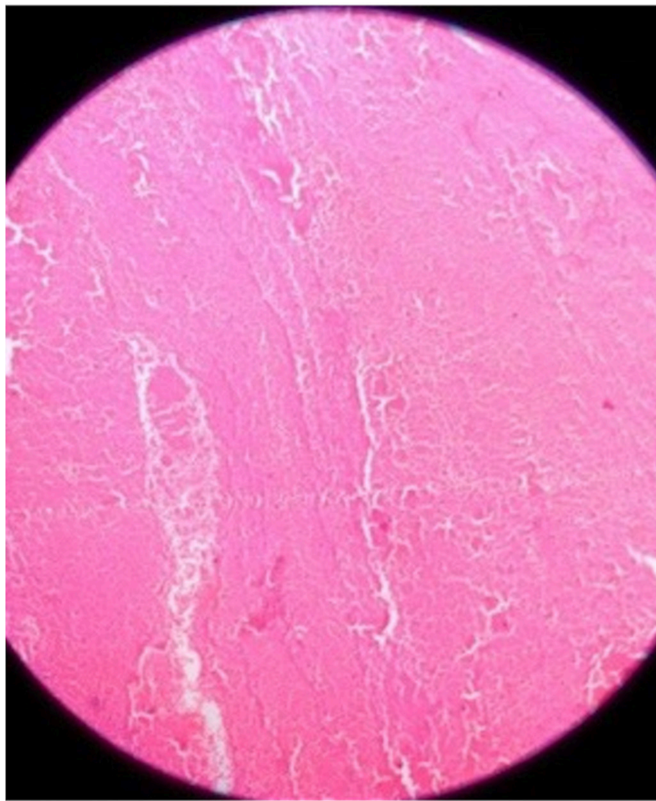

Figure 3 Histopathological slide revealing benign changes with smooth muscle spindles arranged in whorled and parallel bundles, admixed with inflammatory cells.

ones that exhibit one or none of the traits. In this reference, our case describes a large mass of >5 cm. However, it is still suggestive of a benign aetiology. There is a <1% risk of malignant transformation. On MRI, the malignant growth would show low-intensity signals on t2-weighted scans. The mainstay of treatment is surgical excision. Evidence supports the excision of surrounding minor amount of normal tissue that reduces risk of recurrence.[2]

Learning points

- ► Vulvar leiomyoma is a rare and benign lesion that is often misdiagnosed as a Bartholin cyst preoperatively. Usually, they are considered to be masquerading in nature.
- ► Gross appearance, histopathological analysis and MRI findings help to differentiate between benign and malignant vulvar leiomyomas.
- ► The mainstay of management for a benign lesion is surgical excision.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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