

Vulvar Leiomyoma: A Rare Clinical Entity

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Summary

Among all benign tumors, leiomyomas which occur most common in uterus, account for about 3.8%. Vulvar localization of leiomyomas is very rare with only 160 cases reported so far. Here we present a case of vulvar leiomyoma and discuss the diagnosis and management of this disease. A 36-year-old female patient with no medical history, had a 11 x 8 cm mass in the left labia majora causing a sensation of heaviness at the perineum which was aggravated by weight lifting. She underwent a complete surgical excision of the mass. On pathological examination it was diagnosed as a leiomyoma. Vulvar leiomyoma is a rare benign tumor. The diagnosis is made only postoperatively after resection of the mass. The treatment is essentially based on total excision of the mass with a good prognosis.

Keywords: Vulva, leiomyoma

Introduction

Tumors which involve vulva can be benign, premalignant, and malignant tumors. Among soft tissue tumors, leiomyomas account for about 3.8%.¹ The tumors are supposed to arise from smooth muscle present in blood vessels, erectile tissue or remnant of the round ligament in the labia majora.² Commonest site of its occurrence is the uterus.³ Bartholin's cyst is the most common differential diagnosis for unilateral vulvar swelling in the reproductive age women,⁴ whereas leiomyomas of the vulva are particularly rare. Other differential diagnosis are unilateral inguinal hernia, fibroma, lymphangioma and angiofibrosarcoma. In this case report, we are presenting a case of vulvar leiomyoma with regard to its diagnosis and management.

Case Presentation

A 36-year-old female patient, regularly menstruating para 3 live birth 3, with no significant medical history, presented with vulvar mass since 2 months with a sensation of heaviness at the perineum, aggravated by weight lifting. Patient had no history of fever, abdominal symptoms, vaginal discharge. On vulvar examination, there was a firm mass measuring 11 x 8cm located on the left labia majora. On Per Speculum, cervix, vagina were healthy, on bimanual examination, uterus was normal in size [Figure 1]. On MRI, the mass appeared as well defined, encapsulated, with peripheral hyperintensity and

central hypointensity on T2w image. The lesion showed intense heterogenous post contrast enhancement, with round ligament appearing bulky with multiple prominent vessels. On ultrasound of pelvis and groin there was no prominent inguinal lymphadenopathy and no evidence of bowel loops in the mass. Preoperatively provisional diagnosis was angiofibrosarcoma as suggested by MRI. The patient had a surgical excision of the vulvar mass under spinal anesthesia. Histopathology revealed a proliferation of smooth muscle fibers without atypical mitosis, necrosis and pleomorphism. All the cutaneous margins were free of tumor. Morphological analysis concluded it to be a vulvar leiomyoma. No IHC was kept as histopathology clearly revealed smooth muscle tumor.

Discussion

Leiomyomas are smooth muscle benign tumors. Uterus is the most common site, while other ectopic sites being ovaries, urethra, vulva, bladder, peritoneum and retroperitoneum.⁵ Till now only 160 cases have been reported which support its rarity.¹ Twenty-five cases were reported by Nielsen et al.⁶ In most of the cases its diagnosis is made postoperatively. Preoperatively, it can be confused with a Bartholin's cyst.¹ Some features that support the diagnosis Bartholin cyst are everted labia minora and cystic in consistency of the swelling; however, finding inverted labia minora and firm in consistency of swelling support the diagnosis of vulvar leiomyoma.⁴ There are various presenting symptoms, most common being, painless mass, but can present with pain, erythema or itching.⁶

A study on 95 patients with cutaneous leiomyomas, by Kurdi et al, only 4.2% has vulvar involvement.¹ Leiomyoma of the vulva can occur at any age.⁷ In a study by Nielsen et al of 25 patients with vulvar leiomyoma, the mean age of presentation was 37 years,⁶ whereas the mean age in a study by Katenkamp et al on 21 patients, was 41 years.⁷ Indexed patient was 36 years of age with a tumor diameter being 7 x 6 x 5 cm. On histopathological

examination, vulvar leiomyomas are well circumscribed tumors made up of spindle cells in a myxoid stroma without cytologic atypia and are positive for ER PR on immunohistochemistry. Due to enhanced soft tissue differentiation, MRI is considered as the most appropriate imaging tool to differentiate between a malignant and benign tumor.⁸ Myomas appear as hypointense to isointense on both T1 and T2w images, with homogenous enhancement with contrast.⁸ In our case, MRI showed a well-defined, encapsulated lesion with central hypointensity and peripheral hyperintensity on T2w image. The lesion showed intense heterogenous post contrast enhancement, with the round ligament appearing bulky with multiple prominent vessels. Differential diagnosis in this case was Bartholin cyst, angiomyolipofibroma and fibroma.

Surgical excision is the mainstay in the management of a vulvar leiomyoma, with final diagnosis being after histopathological confirmation.⁴ Only 1 out of 25 patients had recurrence of the vulvar leiomyoma after 10 years in the study by Nielsen et al,⁶ hence close long term follow-up is advisable. There is no role of adjuvant therapy as it is benign condition, all margins were negative.

Conclusion

The diagnosis of a vulvar leiomyoma is often made only after the histopathological confirmation of the resected mass. Ultrasound or MRI may aid in the diagnosis of these extrauterine leiomyomas, which present as a rare entity. Bartholin's cyst, lymphangioma, fibroma, sarcoma, or neurogenic tumor should be considered as the differential diagnosis when treating such tumors. The mainstay of the treatment include surgical excision and long-term follow up.

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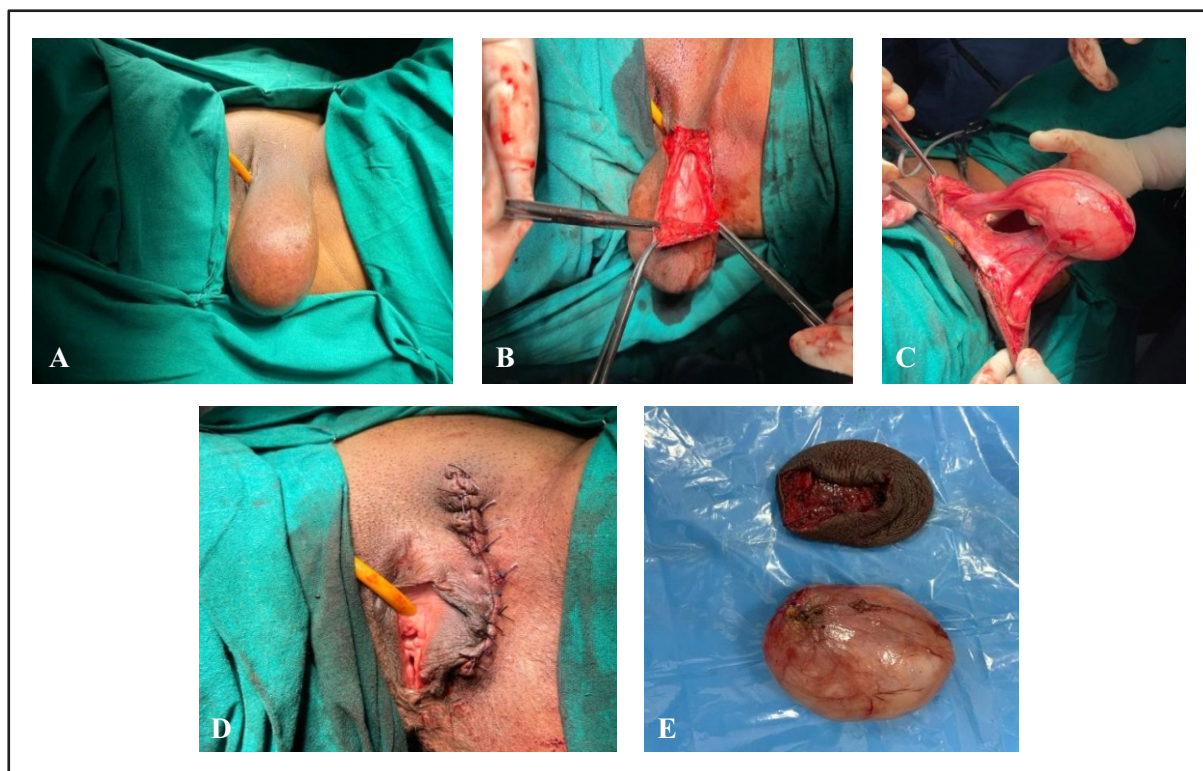


Figure 1: A to D: Intraoperative images of the vulvar fibroid. Notice the pedunculated mass with lax skin wall. E: vulvar mass with covering skin.

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