

DOI: <http://dx.doi.org/10.18203/2320-1770.ijrcog20180929>

Case Report

Leiomyoma of the vulva: a diagnostic challenge

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Received: 25 December 2017

Accepted: 24 January 2018

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ABSTRACT

Smooth muscle tumours of vulva are rare and therefore can be missed clinically. Our patient, 48-year-old lady presented with lump in the left vulva that was clinically diagnosed as Bartholin duct cyst. The lumpectomy was done under local anaesthesia and the lesion was sent for pathological examination. On gross examination the mass was 3.5 cm in diameter. The microscopic examination revealed the lump to be leiomyoma and no necrosis or atypia was present. The final diagnosis of “vulval leiomyoma” was given. Postoperative period was uneventful, and the patient is on regular follow up and there has been no recurrence. Leiomyoma should be kept as a differential diagnosis when a lady presents in late reproductive age group with unilateral swelling in vulvar region which is firm in consistency and the lump must be sent for histopathological examination for definitive diagnosis and rule out malignancy. The pathologists play a critical role in recognition and management of smooth muscle tumors of the vulva and to rule out leiomyosarcoma.

Keywords: Bartholin cyst, Leiomyoma, Vulva

INTRODUCTION

Tumors of the vulva are rare and smooth muscle tumors of the vulva are rarer therefore can be misdiagnosed clinically. The extrauterine leiomyomas are extremely uncommon therefore pose a greater diagnostic challenge.¹ In literature there are limited reports of Leiomyoma vulva.²⁻⁵

Identifying leiomyosarcoma in this region is difficult. It is important to diagnose malignancy due to the risk of recurrence as patients may need radiation and/or chemotherapy in addition to wide local excision and appropriate follow up.

We present an interesting case of leiomyoma of vulva, masqueraded as Bartholin cyst clinically. This case report emphasizes the role of pathologist in diagnosis and management of such cases.

CASE REPORT

48-year-old P2L2 lady, both normal vaginal deliveries, presented with lump in the left vulva which gradually progressed in size over the past 5 years. The lump was not associated with pain or any discomfort locally. Her menses were regular with normal flow and duration. No history of significant weight loss or any other medical or surgical illness. Clinically the lump was felt medial to the left labia minora and measured 3.5 cm diameter. The lump was mobile and non-tender. No erythema, bleeding or local discharge was noted.

The routine investigations of the patient were normal. Ultrasonography of pelvis revealed large fibroid located fundoposteriorly in the uterus measuring 8 × 7 cm. The endometrial lining and both the ovaries were normal. Clinical diagnosis of Bartholin cyst was made.

An elective excision (cystectomy) of the vulval mass was performed under local anaesthesia. The mass was non-adherent to nearby structures including levator muscles, rectum, vagina, or pubic ramus. The mass was removed entirely. Grossly the tissue was globular with smooth outer surface and measured $3.5 \times 3 \times 2.5$ cm. The cut surface showed pearly white solid tumour with whorled appearance (Figure 1).

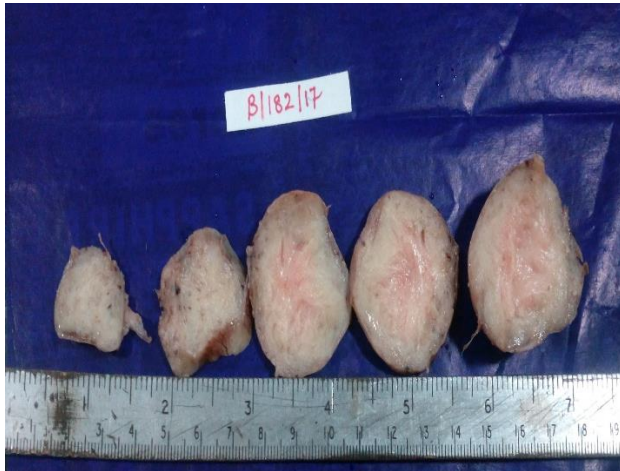


Figure 1: Gross appearance of leiomyoma showing serial cut sections which reveal grayish white lesion with whorled cut surface.

No necrosis, haemorrhage or cystic structure was noted grossly. Microscopic examination revealed well-circumscribed tumour comprising of interweaving fascicles of bland spindled cells with cigar shaped nucleus.

The mitotic figures were less than two mitoses per 10 high-power fields (HPF), and no necrosis was present (Figure 2). The final diagnosis of "vulval leiomyoma" was given. Postoperative period was uneventful. Patient is on regular follow up and there has been no recurrence.

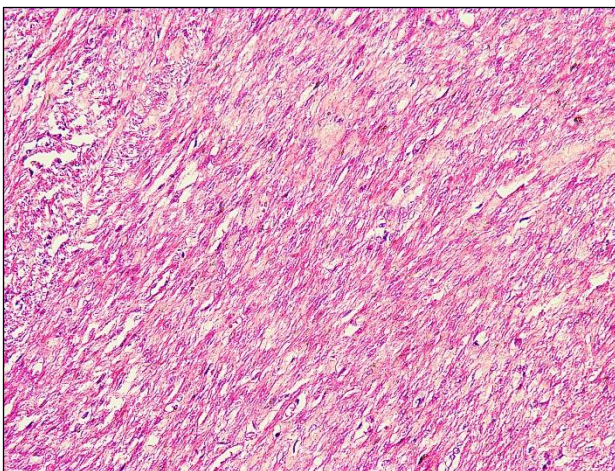


Figure 2: Photomicrograph of Leiomyoma vulva. (H&E x 200)

DISCUSSION

Leiomyomas, are benign soft tissue tumors that arise from smooth muscle, account for approximately 3.8 % of all benign soft tissue tumors.⁶ Uterine leiomyomas affect 20%-30% of women older than 35 years.⁷ The tumors are thought to originate from smooth muscle cells within blood vessel walls or the erectile tissue, the round ligament.⁸ External genital leiomyomas are extremely rare and usually mimic a Bartholin gland cyst.² This case is unique as fewer than 160 cases of smooth muscle tumours of the vulva have been reported in the literature.²⁻⁵

Differentiating leiomyoma from atypical leiomyoma and leiomyosarcoma is challenging. Tavassoli and Norris have proposed the following diagnostic criteria for leiomyosarcoma:⁹

- More than 5 cm in greatest dimension
- Infiltrative margins
- More than 5 mitotic figures per 10 HPF

Nielsen et al studied clinicopathological features of 25 cases of smooth muscle tumours of vulva comprising of leiomyoma and five leiomyosarcoma.¹⁰ He added a fourth criterion, that is presence of moderate to severe cytological atypia. Nielsen et al suggested that if at least three of these criteria are met, the diagnosis is leiomyosarcoma is given. If only two criteria are met, a diagnosis of atypical leiomyoma is warranted, and if only one criterion is met, the diagnosis is leiomyoma.¹⁰ Furthermore Nucci and Fletcher reported the presence of coagulative necrosis to be a feature of malignancy and its presence in combination with any of the four abovementioned criteria should raise the possibility of leiomyosarcoma.¹¹

The treatment for leiomyosarcoma is wide local excision with negative margins and with or without adjuvant therapy.^{10,12} Lumpectomy remains the treatment for the leiomyoma and atypical leiomyoma followed by long-term, careful follow-up.¹⁰ In the present case none of the atypical features were found and it was diagnosed as benign vulval leiomyoma on histopathology.

The differential diagnoses of vulval swelling other than Bartholin cyst are fibroma, lipoma, lymphangiomas, soft tissue sarcoma and neurogenic tumors. So, when a firm vulval swelling is found, leiomyoma should be kept as one of the differential diagnosis and biopsy must be done to exclude leiomyosarcoma.

CONCLUSION

Leiomyoma of vulva is a very rare condition and is usually misinterpreted as Bartholin cyst. It should be kept as a differential diagnosis when a lady in late reproductive age group presents with unilateral swelling in vulvar region which is firm in consistency.

Distinguishing between benign and malignant forms of vulvar leiomyoma is a great diagnostic challenge, excision biopsy and histopathological examination should be done to rule out malignancy.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Saxena S, Goel S, Sen AD. Leiomyoma of the vulva: a diagnostic challenge. *Int J Reprod Contracept Obstet Gynecol* 2018;7:1246-8.