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VULVAR LEIOMYOMA :AN UNUSUAL ENTITY AT A RARE ANATOMICAL SITE

[1]Mohtashim Idrees Akhtar, [2]Dr. Naushaba Tazeen, [3]Dr. Idrees Akhtar Afroze

[4]Dr. Atiya Begum,

MBBS 2nd-year student, Assistant Professor, , Professor and HOD, Professor

Department of Pathology

Deccan College of Medical Sciences, Hyderabad, Telangana

Abstract :

Vulvar leiomyoma is a rare soft tissue tumor, accounting for only 0.03% of all gynecological tumors, which often leads to misdiagnosis. Leiomyoma is a type of smooth muscle tumor that primarily occurs in the uterus and is commonly known as a fibroid. However, extrauterine fibroids are quite rare and can be found in various locations, including the vulva, vagina, ovaries, urinary bladder, urethra, round ligaments, uterosacral ligaments, inguinal canal, retroperitoneum, sinonasal cavities, and even the kidneys.Vulvar leiomyoma typically affects women of reproductive age and can mimic conditions such as Bartholin's cyst or abscess. Histologically, vulvar leiomyomas generally exhibit spindle-shaped cells, though other variants, such as the epithelioid subtype, have also been reported.

In this report, we present a case of vulvar leiomyoma that was misdiagnosed as a Bartholin's cyst before surgery.

Keywords :

Vulvar leiomyoma, bartholin's cyst, misdiagnosis

I.Introduction:

Leiomyomas are well-defined, benign soft tissue tumors of mesenchymal origin, with the uterus being the most common site of occurrence [1]. However, extrauterine leiomyomas are extremely rare and can develop in areas such as the vulva, vagina, ovaries, urinary bladder, urethra, round ligaments, uterosacral ligaments, inguinal canal, retroperitoneum, sinonasal cavities, and kidneys [2].

Vulvar leiomyomas are particularly uncommon, accounting for only 0.03% of all gynecological tumors and 0.07% of all vulvar neoplasms [3]. They typically affect females of

reproductive age and can be asymptomatic [4, 5]. When symptoms do occur, they may include painless swelling of the vulva, often accompanied by erythema or pruritus. Due to their rare occurrence, these tumors can easily be misdiagnosed or go untreated.

Vulvar leiomyomas can mimic Bartholin cysts or abscesses, which creates significant diagnostic challenges. Another difficulty in diagnosing vulvar masses lies in differentiating between benign and malignant forms, as many have a similar appearance. In cases where a similar lesion is observed, transperitoneal ultrasound and magnetic resonance imaging can assist in making a diagnosis [12].

In India, there have been only a few hundred reported cases of vulvar leiomyoma.

Case report

A 38-year-old married female, P4L4 and sexually active presented to the clinic with complaints of painless vulvar swelling for 6 years which was gradually increasing in size. There was no history of dysfunctional uterine bleeding, no history of vaginal bleeding, fever, weight loss, or bowel or bladder complaints. Her past medical and surgical history were unremarkable.

General examination was unremarkable. Local examination revealed a vulval firm solid mass measuring 8cm x 6cm. It was non-tender and not adherent to the overlying skin or the surrounding structures. There was no associated inguinal lymphadenopathy. Their family history was unremarkable.

Transperineal ultrasonography revealed a solid mass measuring 6cm x 8cm x 6cm with normal internal genitalia. The mass was misdiagnosed as a bartholins cyst. Surgical excision was performed for the same. The excision at the mucocutaneous junction showed a firm fleshy well-defined mass of 8cm x 6cm x 6cm. The mass was enucleated as a whole and sent for histopathological examination, postoperative recovery was complete without any complications.

Histopathological examination was performed on a single grayish-white soft tissue mass measuring 6cm x 8cm x 8cm. The cut section of the specimen was grayish-white as well. Sections studied microscopically show an encapsulated mass with intersecting fascicles of plump spindle cells with eosinophilic cytoplasm, oval nuclei with prominent nucleoli having benign features, and there is no evidence of mitotic activity. Intervening tissue showing intense hyaline change.

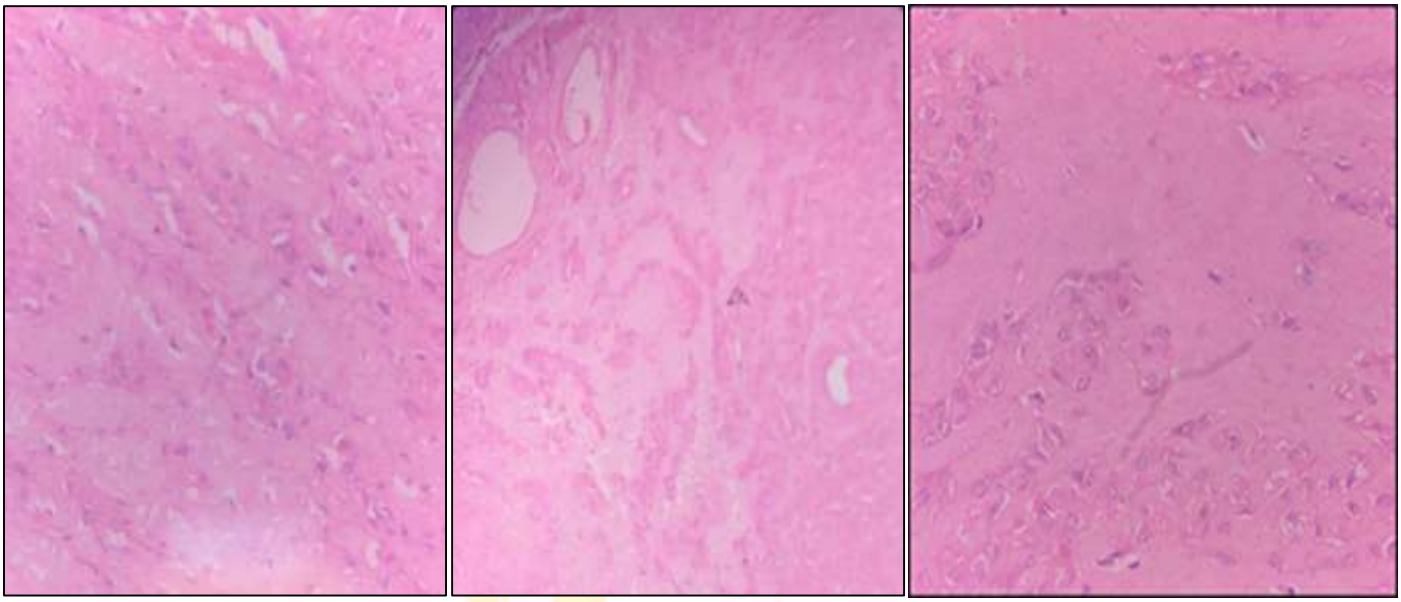

Figure 1 : Microscopy(10x/40x) - Intersecting fascicles of plump spindle cells with eosinophilic cytoplasm, oval nuclei with prominent nucleoli having benign features. Intervening tissue showing intense hyaline change.

Discussion:

Leiomyomas of the female genital tract are primarily found in the uterus, followed by the cervix, round ligament, uterosacral ligament, ovary, and inguinal canal. Vulvar leiomyomas, however, are extraordinarily rare, with fewer than 300 cases reported since the first one was identified in 1908 [6, 7]. The infrequency of these lesions makes them unique. They originate from smooth muscle cells within erectile tissue, blood vessel walls, the round ligament, erector pili muscle, and stem cells located in the Bartholin's gland [8].

Clinically, vulvar leiomyomas are often misdiagnosed as Bartholin's cysts or abscesses before surgery. Surgical excision, including some surrounding normal tissue, is the best treatment option. Patients typically present with a single, painless swelling that increases in size over time, potentially causing difficulties in sitting, walking, or engaging in sexual activity. The size of the swelling can range from 0.5 cm to 15 cm [9]. For instance, one patient presented with an asymptomatic swelling that gradually increased to a size of 6 cm x 8 cm x 8 cm. Less common symptoms may include pain resulting from local irritation of peripheral nerves, pruritus, erythema, and occasional compression of the urinary bladder or rectum [3]. Vulvar leiomyomas generally feel firm but can become soft and cystic due to degenerative changes, complicating diagnosis due to their nonspecific presentation.

Often, these tumors are misdiagnosed as Bartholin's cysts because of topography of the bartholins gland or as, lipomas, fibromas, schwannomas, aggressive angiomyxomas, hemangiomas, leiomyosarcomas, dermatofibrosarcomas. Clinical findings suggestive of a benign solid lesion include everted labia minora and a firm, nontender, mobile, regular-shaped swelling [9, 10, 11, 12].

Diagnostic options include transperineal ultrasonography and magnetic resonance imaging (MRI). Differentiating between benign and malignant variants poses a challenge. Nielsen et al. proposed a criterion for distinguishing these based on four features: the widest dimension greater than 5 cm, more than 5 mitotic figures per 10 high power fields, and moderate to severe

cellular atypia. If three of these findings are present, the tumor can be considered a neoplasm. Benign but atypical leiomyomas exhibit only two characteristics, while benign leiomyomas show one or none. In the case discussed, no findings suggested malignancy, indicating a benign leiomyoma. MRI findings of malignant variants typically show low-intensity signals on T-2 weighted scans, unlike normal smooth muscle cells [12]. Histopathological examination of surgically excised specimens is the gold standard for final diagnosis and ruling out malignant variants.

Vulvar leiomyomas may test positive for estrogen and progesterone receptors on immunohistochemistry. Treatment options include enucleation and wide local excision, with some normal surrounding tissue removed to reduce the risk of recurrence. Selective receptor therapy can be administered as an adjunct to surgical procedures for tumors that are positive for estrogen and progesterone receptors [12]. There is a notable chance of recurrence and the need for regular long-term follow-up after treatment, as emphasized in studies by Nielsen et al. and Kothandaraman et al [2, 14].

Conclusion

Vulvar leiomyomas are rare, benign tumors that can be challenging to diagnose due to their infrequent presentation and resemblance to other vulvar masses, such as Bartholin's cysts. This case highlights the importance of considering vulvar leiomyomas in the differential diagnosis of vulvar masses, especially in women presenting with painless, firm, and gradually enlarging swellings. Imaging modalities like transperineal ultrasonography and MRI, alongside histopathological examination, play a critical role in the accurate diagnosis and distinction of benign from malignant variants. Surgical excision remains the preferred treatment for vulvar leiomyomas, with wide local excision recommended to prevent recurrence. Given the possibility of recurrence and the slow but progressive nature of these tumors, ongoing follow-up is essential. The potential presence of hormone receptors also opens avenues for selective receptor-based therapies, providing adjunctive options for cases where surgical excision may not be definitive. This case underscores the need for awareness of this rare entity to avoid misdiagnosis and ensure optimal patient outcomes. It is only a pathologist who can give the correct diagnosis and guide the clinician in this matter.

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