A rare epithelioid leiomyoma of the vagina with sections of myxoid degeneration

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ABSTRACT

Leiomyomas are common benign tumors in the uterus. Leiomyomas of the vagina are rare, and the epithelioid leiomyoma of the vagina is an extremely rare benign tumor. They are often accompanied by different symptoms. Here, I present the case of a 42-year-old woman, with local discomfort and dysuria. Enucleation of the tumor through a vaginal approach was done successfully without urethra injury as a healing method.

Keywords: *Leiomyoma, Misdiagnose, Vagina.*

Smooth muscle tumors of the vagina are uncommon entities and this rarity could lead to misdiagnosis and go wrong with cysts or abscess.Leiomyomas of the vagina are very rare; approximately 330 cases have been reported in the literature so far [1] and only three were of epithelioid leiomyomas [2]. Usually, they arise from the anterior vaginal wall and depending on the size and location, they may cause varied clinical presentations, such as discomfort, pain, or dysuria [1, 2]. They have a submucosal or a pedunculated growth pattern from the anterior vaginal wall into the vaginal cavity [2].

Here, I report for the first time, a case of an epithelioid leiomyoma with myxoid degeneration, misdiagnosed as vaginal cyst preoperatively but histopathologically confirmed as Epithelioid leiomyoma with myxoid degeneration. This case sheds light on tumor pathology in a woman‘s reproductive system which must be borne in mind from urologists, gynecologists, and pathologists.

CASE REPORT

A 42-year-old woman was admitted to our hospital for the complaints of dysuria and sometimes incontinency. There were no accompanying diseases and no clinical findings revealed pathological abnormalities. The couple announces two pregnancies with two live births children. On systemic examination, the vitals were stable. On Gynecological examination, a round, cystic mass, measuring approximately 3 cm in diameter, was palpable between the urinary meatus and front wall of the vagina. Her cervix has a whitish portion and there were no changes in the uterine body.

Laboratory findings revealed that blood smear and biochemistry were within normal limits. Serum CA-125 in a norm (15.30 U / ml). Ultrasonography examination revealed a soft elastic mass of approximately 3 × 3.5 cm on the front wall of the vagina with no obvious limits. Uterus and adnexa were within normal parameters. Because of the location, laboratory findings, and ultrasonography, a vaginal cyst was considered as the provisional diagnosis.

Surgical excision was done through the vaginal route. A Foley catheter was introduced in the urethra for protecting the latter during the operation. A cross-section was performed on the anterior vaginal wall, whereby a 3 cm soft encapsulated tumor was present within the vaginal wall. There was no relationship between the tumor and the urethra. The tumor was removed completely and sent for histology. The vagina was repaired by the two-layer closure procedure.

Macroscopically, the tumor was 3 × 3.5 cm. The cut was soft elastic and the cutting surface waspale brownish homogeneous with a visible small cyst at one pole (Fig. 1). Histologically, tumor was made up of bundles of elongated spindle cells with abundant eosinophilic cytoplasm and small nuclei without atypia. The stoma was fibrous with thin-walled vessels, edema, and focal myxoid degeneration (Fig. 2a,b). In large sections, the tumor

Figure 1: Macroscopic picture: cut surfaces of the tumor.

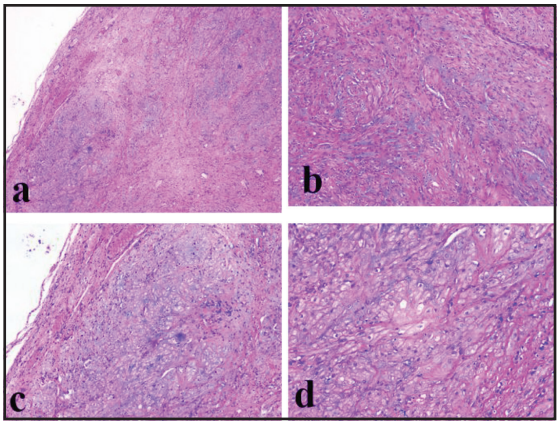

Figure 2: Histology (H&E): (a)Bundles of spindle cells and nuclei without atypia, fibrous stroma with thin-walled vessels, edema, and focal myxoid degeneration (enlargement ×50); (b) bundles of elongated spindle cells with eosinophilic cytoplasm, focal myxoid degeneration (enlargement ×100); (c) In large sections, the tumor showed epithelioid spindle-cell growth without necrosis (enlargement ×100); (d) Nuclear pleomorphism was mild, and mitotic activity was scarce (enlargement ×200).

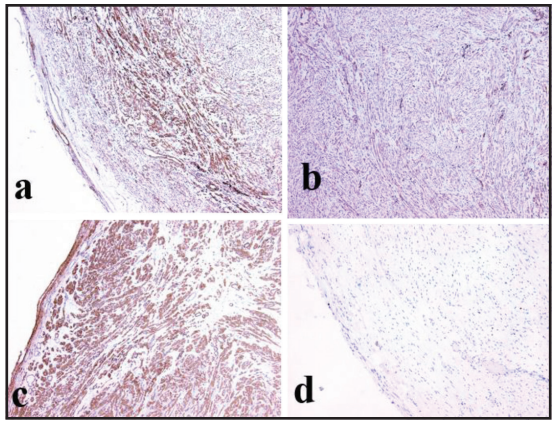

Figure 3: Immunohistochemistry: Positive expression of(a) Vimentin (enlargement x50); (b) Desmin (enlargement x50); (c) SMA (enlargement x50) and (d) Ki 67 not more than5% (enlargement x100).

showed epithelioid short spindle-cell growth without necrosis. Nuclear pleomorphism was mild, and mitotic activity was scarce (Fig. 2c,d). Immunohistochemistry (Dako, Glostrup, Denmark) was done and the tumor was positive for Vimentin, Desmin, SMA (Fig. 3a,b,c) and negative for p53, HMB45, S100, CD34 and Ki67 prolific activity, not more than 5% (Fig. 3d).

On the basis of the above-mentioned investigations, a diagnosis of the epithelioid leiomyoma of the vagina with myxoid degeneration was made. After the operative period, the urine runs smoothly and without complications. Urinary disorders recovered one week after the surgery. At the check-ups by a gynecologist in the first and sixth months after surgery, no recurrence was detected and the laboratory results showed that blood count and biochemistry were within normal limits.

DISCUSSION

Vaginal leiomyomas are rare. Only about 330 cases have been reported since the first detected case back in 1733 by Denys de Leyden [3] and only three reports of the Epithelioid leiomyoma of the vagina are found in PubMed [2]. Our practice encountered the first reported case of vaginal Epithelioid leiomyoma with myxoid degeneration. Vaginal leiomyomata are benign mesenchymal tumors of the vaginal wall that arise from smooth muscle elements and are uncommon. These masses may occur anywhere within the vagina and vary from 0.5–15 cm in diameter. They commonly present as a smooth, firm, round mass on the anterior vaginal wall. Sometimes, they may be misdiagnosed as urethral diverticulum or paraurethral cyst [1]. In the present case, the clinical diagnosis was for a vaginal cyst.

Vaginal leiomyomas usually develop in women between the age of 35 and 50 [4] but in our case, it was seen at age 42. These lesions are usually estrogen-dependent and can grow rapidly during pregnancy or regress after menopause [4]. Before surgery in low resources setting, an appropriate pelvic examination for localization and exclusion of pelvic organ prolapse and ultrasound may help to estimate the extent because we didn't have sophisticated equipment like magnetic resonance imaging (MRI) and urethrocystoscopy.

In the case reported, due to its soft consistency, the tumor has misled the gynecologist who suggests that the cyst is involved. The unusual macroscopic and microscopic look was a challenge for us. It is estimated from routine histology that it is not a cyst, but a benign mesenchymal tumor. The presence of bundles of elongated spindle cells without atypia and fibrous stroma directs to leiomyoma, but extensive epitheloid cells with abundant eosinophilic cytoplasm and the edema, myxoid degeneration areas they are disturbing. This forced us to appoint immunohistochemistry to determine the immunophenotype of the tumor.

The immunohistochemical staining pattern was positive for vimentin, desmin, alpha-smooth muscle actin, which supports the morphological diagnosis for mesenchymal tumor (leiomyoma). The tumor was negative for HMB45, and S100, which excluded the diagnosis of melanoma.Negative HMB45 excludes the possibility of a perivascular epithelioid cell tumor. Negative p53 and low proliferative activity studied with Ki67, as well as the presence of fewer than 2 mitoses in 10 visual fields at x400 magnification, do not support leiomyosarcoma [5]. CD34 expression was negative, which excluded a solitary fibrous tumor.

The pathological diagnosis was an epithelioid leiomyoma with sections of myxoid degeneration. This case is an example of unusual localization of a rare type of leiomyoma in the female reproductive system. Up to now, three cases of epithelioid leiomyoma of the vagina [2] have been reported. The case reported is the first to epithelioid leiomyoma with sections of myxoid degeneration of the vagina. Six months after surgery, the woman was well with normal laboratory results and no recurrence.

CONCLUSION

This is an example that despite the limited resources, good clinical preparation is beneficial to the patient. From the present case and the review of the literature, it is confirmed that the final diagnosis is morphological.

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