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CASE REPORT

IMT of Vulvovaginal Region: A Rare Case Report with Recurrence

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Introduction

A wide variety of soft tissue tumors (STS) can occur at vulvovaginal region of a woman. STS accounts approximately 1% of all adult tumors. Inflammatory myofibroblastic tumor is an even rarer STS. These are unique subset of neoplasms which comprises of fibroepithelial stromal polyp, myofibroblastoma, angiofibroma angiomyofibroblastoma and aggressive angiomyxoma. Most of them arise from either stromal cells or mesenchyme cells of endocervix to vulva. Inflammatory myofibroblastic tumor (IMT) of vulvovaginal region is even a rarer spindle cell neoplasm with very low malignant potential [1]. It is mesenchymal neoplasm composed of mainly proliferative myofibroblasts and fibroblasts accompanied by a mixed population of inflammatory cells including plasma cells, lymphocytes and eosinophils. This tumor was first described in lung, but is known to occur in many extrapulmonary sites including female genital organs, most commonly the uterus. The occurrence of an inflammatory myofibroblastic tumor is extremely rare in the female genital tract. The tumor is more common in children with no predilection for sex, but in adult age, it is more common in women. Symptoms and treatment are heterogeneous; mainly depend on the localization of the tumor [2, 3]. IMT of vulvovaginal region presents in adult women most commonly in their 50 s, as a nodular or polypoidal painless growth. Although they manifest as benign tumor of vulva, vagina or cervix, they locally behave as aggressive and recurrent tumors. There is limited literature available regarding clinical presentation and management of these tumors. Consequently, definitive diagnosis is often challenging for pathologists because of rarity of tumor. It is further complicated by either limited biopsy specimen for evaluation or unknown information about the vague clinical presentation. Most of time diagnosis is only made after histopathological report and by immunohistochemistry. Histologically, IMTs are composed mainly of fascicles of myofibroblasts and fibroblasts spindle cells. These tumors are positive for smooth muscle actin, vimentin, desmin and b-catenin. Desmin is positive in 75–100% of cases, ER and PR are positive in 80–100%, CD34 is positive in 50–85%, and smooth muscle actin is positive in 0–45% [4]. ALK immunohistochemistry is positive in 50–60% of cases and generally correlates with rearrangement of ALK. We here report a rare case of vaginal myofibroblastic tumor with recurrence seen. Final diagnosis was made with histopathology and immunohistochemistry report.

Case Report

A 46-year P3L3 lady visited to gynecological oncology OPD with history of operation for vaginal wall growth two times at private local practitioner. Histopathology report presented by her showed grossly as 7 × 4.5x2.5 cm globular tissue. On cut section, it showed congested and myxoid areas. Final HPE report showed spindle cell neoplasm (Fig. 1).

Her menstrual cycles were regular. On clinical examination, introitus was normal with induration present near posterior fourchette, no any obvious growth found. Cervix was smooth; uterus and B/l adnexa were normal. We reviewed the block of specimen provided by patient at our center where both HPE and IHC were done which confirmed myofibroblastic tumor. IHC was SMA positive, B-catenin positive, CD68 focal positive, CD34-positive around blood vessels and Ki-67 -2% (Fig. 2a, b, c). This lady has been

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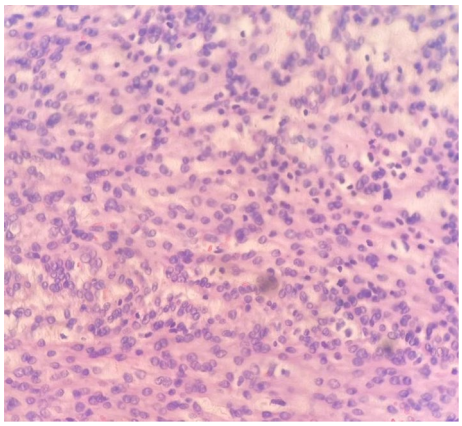

Fig. 1 Myofibroblastic spindle cell proliferation

operated first time in January 2021. 2nd time operation was done by another gynecologist 2 month later. She has been in regular follow-up since April till today in Gynae-oncology department. On her first visit, she was digitally examined both vaginally and per rectally to detect any growth. Per speculum examination was done to know the status of vaginal walls and cervical lips. Perineal ultrasound was performed which did not showed any abnormality. So further no radiological imaging like CT and MRI scan was administered. She is clinically monitored regularly with per speculum examination on her each visit. There is no recurrence of symptoms seen yet.

Discussion

IMT of vulvovaginal region is a comparatively rare enigma that is almost unknown to many gynecologists. Less than 5 cases have been reported so far on IMT of vaginal region. Many cases related to uterine-located IMT have been reported. Our case of vaginal region with recurrence will be the first one to be reported so far. Sometimes IMT is referred to as plasma cell granuloma, or inflammatory pseudo tumor (IPT) [1]. Recently because of cytogenetic clonality, recurrent involvement of chromosomal region, occasional aggressive local behavior and metastasis of the tumor, IMT is considered a neoplasm rather than a post-inflammatory process.

IMT should be differentiated from other tumor-like lesions of the vulvovaginal region such as Bartholin cysts, benign lipoma, fibroepithelial stromal polyps, cellular angiofibromas, carcinoma of cervix, condyloma accuminata, endocervical polyp and pedunculated submucous myoma. Complete surgical resection is the mainstay of the treatment of these tumors. Recurrence rate for IMT is about 25% cases, while distant metastases are seen in only 2%. In our case, we found recurrence due to inadequate tumor negative margin and less practical information to how to handle these tumors. Tumor behavior mostly depends on the size, mitosis, tumor cell necrosis and margin of resection line [3]. Recurrence can be the outcome of incomplete resection of tumor. In some cases of hepatic IMT, spontaneous regression has also been seen thus favoring "wait-and-see" approach as a safe treatment of IMTs for both pediatrics and adult cases. IHC serves as a diagnostic tool for differentiating IMT from other tumors of vulvovagina as seen in our case. Fifty percent of IMT have ALK genetic rearrangement which combines with many different genes to make different ALK proteins. This protein acts as oncogenic trigger. Markers of myofibroblastic differentiation are SMA, desmin and cytokeratins. Ki-67 index is generally seen low in these tumors. IMTs shows negativity with immunostains like S100 protein and myoglobin. There is generally overlapping of IHC markers seen with other varieties of mesenchymal soft tissue tumors. Desmin, SMA and estrogen receptor/progesterone receptor are variably expressed by different subtypes of STS. So histopathology together with IHC complements each other in establishing diagnosis of such an extremely rare variety of tumor as happened in our case.

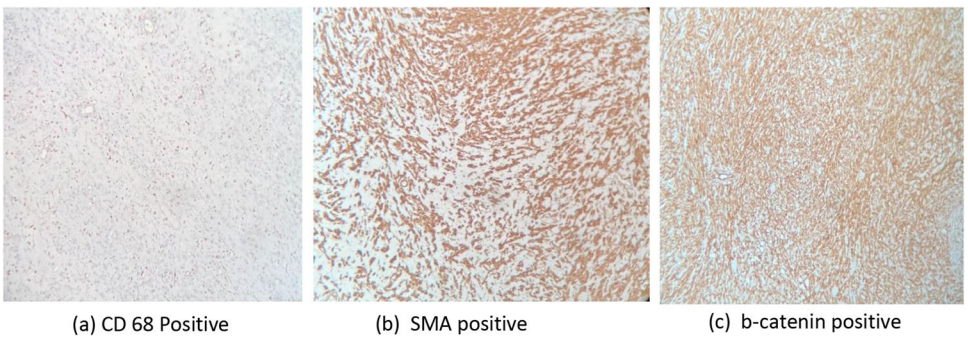

Fig. 2 IHC **a** CD 68 positive, **b** SMA positive, **c** b-catenin positive

Conclusion

The case reported here is extremely uncommon recurrence of IMT of vaginal region. Only few cases have been reported very so far. Although pathology is the gold standard for diagnosis, sometime IHC is essential for the diagnosis of this rarer variety of soft tissue tumors. Due to the low number of published cases, the current incidence of IMT of vulvovaginal origin is difficult to be established. This case will aid in providing practical information for the general gynecologists as to how to handle these uncommon tumors of intermediate malignant potential.

Declarations

Conflict of interest The authors declare no conflicts of interest.

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