CASE REPORT

A rare case of lipoleiomyoma of the vulva: Cytological and immunohistopathological study

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ABSTRACT

In clinical practice, smooth muscle tumors of the vulva are rarely encountered. The histopathological subtype and grading is of utmost importance to the clinician and the patient. Diagnosis is based on clinical presentation, radiological, cyto-histological examination, and recently by application of immunohistochemical (IHC) markers. Leiomyomas occasionally occur with unusual patterns, making their identification more challenging clinically, radiologically, and cyto-histologically. We encountered a case of lipoleiomyoma of the vulva in a 38-year-old female. To the author's knowledge, this unusual variant of smooth muscle tumor, that is, lipoleiomyoma of the vulvar region has not been mentioned in the literature so far. The detailed clinical, cytomorphological, histopathological, and immunohistochemical study was carried out, which has prompted us to report this case.

Key Words: Cytological, fine needle aspiration cytology, immunohistochemical, lipoleiomyoma, vulva

INTRODUCTION

In addition to malignant lesions, benign neoplastic lesions encountered in vulvar region by clinicians are melanocytic nevi, hemangiomas, seborrheic keratosis, hidradenomas, basal cell carcinoma, and unusual tumors such as neurofibromas and syringomas.[1] It is worth noting that not a single case of leiomyoma is mentioned in the above study carried out over a period of 8 years. Smooth muscle tumors (SMT) of the vulva are rare with an incidence varying from 0.07% to 4.2%.[2] The most common type described in vulvar region is leiomyoma showing myxoid and that showing hyaline change.[2] Few single case reports of epithelioid and fibroma like leiomyomas of the vulva have been documented.[3,4] Since the advent of IHC, there have been reports documenting hormonal markers such as estrogen receptors (ER) and progesterone receptors (PR) besides the markers for specific type of tumors based on histopathological examination in lower female genital tract neoplasms. The present case of lipoleiomyoma of the vulvar region is extremely rare. To the author's knowledge, lipoleiomyoma of the vulvar region has not been mentioned in the literature so far. Our experience with the present case adds to a rare variant of leiomyoma arising in the vulvar region, its cyto-histological correlation and significance of immunohistochemical analysis in diagnosis.

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CASE REPORT

A 38-year-old female, gravida 2 and para 2, presented to gynecology outpatient department with a left sided mass in the vulvar region. It was noticed one year back, which was small initially and gradually increased to the present size. Local examination showed 3 × 3 cm, nontender, well defined, soft to firm swelling in the labium majus. Overlying skin was unremarkable. Systemic examination findings were noncontributory. Fine needle aspiration cytology (FNAC) was advised.

FNAC revealed moderately cellular smears composed of tissue fragments of cells with indistinct cell margins, moderate amount of eosinophilic cytoplasm and spindly nuclei of varying size. Few adipocytes were seen entrapped within the spindle cell fragments [Figure 1]. Background was hemorrhagic. Cytological report of benign spindle cell lesion with differential diagnosis of spindle cell lipoma

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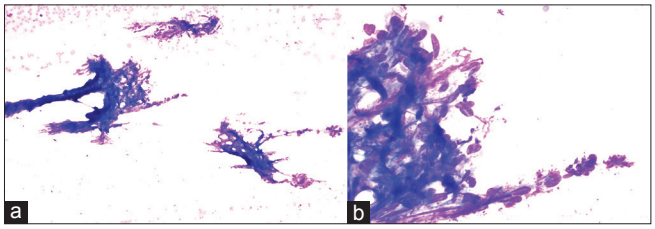

Figure 1: Fine needle aspiration cytology – Tight spindle shaped cells cluster admixed with mature adipocytes. (a) (Leishman ×100), (b) (Leishman ×400)

and leiomyoma was offered. The complete excision of the lesion was advised for histopathological confirmation.

On gross examination, the tumor was ill circumscribed, partly encapsulated, soft to firm measuring 6 × 2.5 × 2 cm. On cut section, the tumor appeared whitish with yellowish and few tiny hemorrhagic areas [Figure 2a].

On microscopic examination, an ill circumscribed tumor mass composed of spindle cells arranged in interlacing fascicles and whorls, regularly interspersed with single and at places collections of mature adipocytes was observed. The spindle cells showed bland oval nuclei and moderate amount of eosinophilic cytoplasm [Figure 2b]. No areas of necrosis and abnormal mitosis were noted. Based on histomorphologic appearance, differential diagnosis of lipoleiomyoma and neurofibroma with lipomatous differentiation was considered and representative sections were subjected for immunohistochemical studies.

IMMUNOHISTOCHEMICAL FINDINGS

Immunohistochemistry was performed with the following panel of antibodies, namely, desmin (clone33, Dako), smooth muscle actin (SMA) (clone 1A4, Novacastra), S-100 (Leica), CD-34 (clone QBEnd/10, Novacastra). Hormonal status markers like ER (clone 6F11, Novacastra) and PR (clone PGR312, Novacastra) were also studied. The tumor cells showed strong cytoplasmic immunoreactivity for desmin [Figure 2c] and SMA. They were nonreactive for CD-34 and S100. Mature adipocytes showed focal nuclear and cytoplasmic immunoreactivity for S-100. Strong nuclear immunoreactivity for ER [Figure 2d] and PR was shown by both spindle cells and adipocytes.

Based on histopathological and IHC studies, final diagnosis of lipoleiomyoma was arrived. Postoperatively the patient is disease free and on follow-up there is no evidence of recurrence for a period of 6 months.

DISCUSSION

SMT of the vulva are rare with an incidence varying from 0.07% to 4.2%.[2] They include a variety of histologic types.

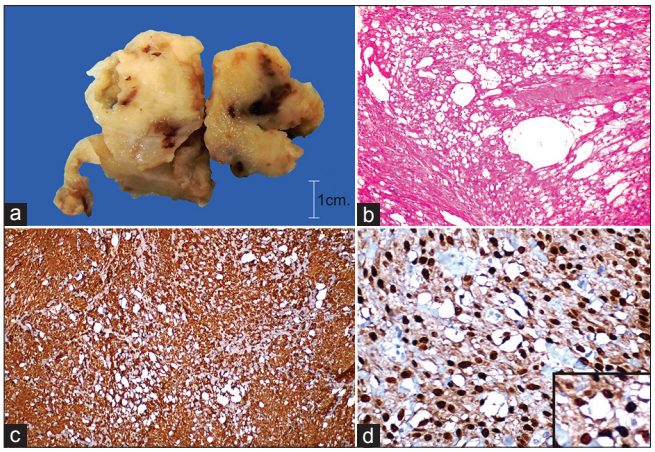

Figure 2: (a) Gross photograph – ill circumscribed tumor appearing whitish with yellowish and few tiny hemorrhagic areas. (b) Photomicrograph of tumor-spindle cells arranged in interlacing fascicles and whorls, regularly interspersed with single and at places collections of mature adipocytes. (H and E, ×400). Immunohistochemical findings: (c) Spindle cells showing strong cytoplasmic immunoreactivity for desmin (×100), (d) Strong nuclear positivity for estrogen receptor (×400), inset showing strong nuclear positivity by adipocytes

Uterus is the most common site of origin of leiomyomas. They arise as proliferation of smooth muscle cells and may develop at any site where such cells are found. Unusual sites of origin include the vulva, ovaries, urinary bladder, and urethra.[5]

Lipoleiomyoma of the uterus is relatively uncommon accounting for 0.03-0.2%.[6] Vulvar leiomyoma showing myxoid and hyaline change have been well documented.[2] It must also be mentioned that few single case reports of epithelioid and fibroma like leiomyomas of the vulva have been reported.[3,4] To the best of our knowledge, lipoleiomyoma of the vulvar region is not mentioned in the literature so far.

With regard to histogenesis of vulvar leiomyoma, it may arise from the network of muscle fibers that lie in the deep dermis or from the smooth muscle of the blood vessel wall of labia majora.[2] The lipomatous component could arise from either fatty metamorphosis of the smooth muscle cells of leiomyoma or lipid metabolic disorders associated with estrogen deficiency in peri- or postmenopausal period promoting intracellular storage of lipids. It has also been documented that fatty tissue is active proliferative tissue and not degenerative change.[7]

Magnetic resonance (MR) imaging is the most useful imaging modality for characterizing these tumors, because, regardless of their anatomic location, classic leiomyomas have signal intensity similar to that of smooth muscle on images obtained with any MR pulse sequence.[5] However, histopathologic analysis is required to confirm

the diagnosis. Radiological investigations were not done in our case.

The clinical differential diagnosis of vulvar tumor includes benign and malignant entities such as bartholin cysts, fibromas, lymphangiomas, soft-tissue sarcomas, and neurogenic tumors.[5] The microscopic differential diagnosis of lipoleiomyomas includes spindle cell lipoma, angiolipoma, angiomyolipoma, and dedifferentiated liposarcoma.

A typical leiomyoma can be diagnosed through examination of hematoxylin and eosin-stained tissue section by an experienced pathologist, but it is sometimes difficult to differentiate an atypical leiomyoma from a spindle cell soft tissue tumor and needs immunohistochemical stain for a confirmatory diagnosis.[2] Lipoleiomyomas are immunoreactive for desmin and SMA, which was also demonstrated in our case. They are negative for S 100 and CD34.[2] Our case showed focal immunoreactivity for S 100. This is attributed to the immunoreactivity showed by the adipocytes.

Immunohistochemically, ER and PR are expressed in 73.7% and 85% of SMTs of vulva, respectively. Absence of ER and PR activity is commonly seen in myxoid leiomyoma suggesting suppression of ER and PR expression in SMTs of vulva.[2] It is worth mentioning that our case showed strong nuclear immunoreactivity for ER and PR not only in smooth muscle cells but also in adipocytes. It also reinforces the fact that the fat is proliferative and not degenerative and is specific for female genital tract fat.[7]

Like SMTs of the uterus, the major diagnostic problem with SMTs of the vulva is distinction between benign and low grade malignant forms. 25% of SMTs of vulva are atypical leiomyomas and leiomyosarcomas. Gunnlauger *et al.*, proposed that the most common finding in SMTs of vulva that recurred, metastasized or both, include a diameter of 5 cm or greater, an infiltrative margin, a mitotic count of 5 or more per 10 high power fields and grade two to three atypia.[8] Other study suggested that the distinction of leiomyosarcomas from leiomyomas is based on the concomitant presence of nuclear atypia, high mitotic index, and zonal necrosis in leiomyosarcomas. In addition, the presence of 10 or more mitotic figures per 10 high-power (×400) fields indicates a malignancy, regardless of cellular atypism. Furthermore, if the tumor contains prominent nuclear atypia or large cells, 5 mitotic figures per 10 high-power fields are sufficient to justify a diagnosis of malignancy.[8] Benign leiomyomas are known to have the potential to recur after many years.[2] Also few cases of leiomyosarcomas have known to arise in uterine lipoleiomyomas.[9] So long-term follow up for local recurrence and malignant transformation is indicated.

Vulval tumors are known to have longest diagnostic delay in gynecological malignancies.[10] Though early diagnosis due to visual inspection is expected, the delay is either due to lack of seeking medical attention by the patient considering the lesion as insignificant or due to nonperformance of local examination by the general practitioner as patient does not come with the symptom of vaginal bleeding. The variability in clinical appearance of vulvar tumors suggests that biopsy confirmation should be obtained on all lesions for which there is the least doubt regarding the diagnosis.[1]

Our experience with present case highlights the rarity of lesion, cyto-histological correlation, and significance of immunohistochemical analysis in diagnosis. It is hoped that the lipoleiomyoma will be kept under differential diagnosis in midlife woman presenting with vulvar mass lesion.

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