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Case Report

Smooth-muscle tumor of vulva with uncertain malignant potential: A case report

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Abstract

Smooth muscle tumors of uncertain malignant potential (STUMP) of vulva are rare and usually misdiagnosed as Bartholin gland cyst or abscess. In this paper, a 33-years-old woman who was misdiagnosed as aggressive angiomyxoma was presented. The patient was presented with 8x9 cm mass on the left vulvovaginal wall extending into retroperitoneal abdominal space. On pelvic magnetic resonance imaging, there was a 7x9 cm lesion between posterolateral to the bladder and ishio-anal fossa. Biopsy was taken from the lesion revealing aggressive angiomyxoma. Wide local excision of the mass was done extending posteriorly to the ischio-anal fossa, superiorly up to clitoris and left vesical space, medially vaginal wall and laterally up to the ischial tuberosity. The pathology result was reported as STUMP with tumor free margins. No adjuvant therapy was administered and the patient had an uneventful recovery, with no recurrence so far for 12 months. Although usually benign, STUMP requires thorough pathological and radiological workup to rule out malignant features and other close differentials. Early aggressive resection is recommended due to destructive nature of this tumor on adjacent tissues. Although the experience with follow-up of this tumor is scarce, the prognosis depends on complete surgical resection and lack of pathological features of malignancy.

Key Words:

Smooth muscle tumor of uncertain malignant potential, vulva, aggressive angiomyxoma, recurrence

Introduction

Leiomyomas account for approximately 3.8% of all benign soft tissue tumors [1]. They can arise anywhere in the body where smooth muscle is present, the most common site is the uterine myometrium [2]. Smooth muscle tumors of the uterus may be benign or malignant on histologic evaluation. Three recognized features of malignant smooth muscle tumors of the uterus are moderate to severe cytological atypia, a mitotic count of ≥10 mitotic figures (MF) per 10 high power fields (HPF) and tumor necrosis. If a tumor meets 2 of the 3 features, it is diagnosed as a leiomyosarcoma [3]. In contrast, benign leiomyomas of the uterus are defined as smooth muscle tumors with no atypia ≤4 MF per 10 HPF and no tumor cell necrosis. Smooth muscle tumors that do not meet these criteria are diagnosed as smooth muscle tumors of uncertain malignant potential (STUMP) [4]. External genital leiomyomas are rare and usually misdiagnosed as Bartholin gland cyst or abscess. Vulvar smooth muscle tumors occur predominantly in premenopausal women, typically in the fourth and fifth decades. Most are benign or at most, locally recurring .They have different criteria than uterine tumors for distinguishing benign and malign tumors. They are considered to be malignant when they have 3 or all 4 of the following features: (1) size larger than 5 cm, (2) infiltrative margins, (3) moderate to severe cytological atypia, and (4) more than 5 mitoses /10 HPF [4,5]. Smooth muscle tumors can be misdiagnosed as several other conditions such as aggressive angiomyxoma, myxoid liposarcoma, myxoid variant of malignant fibrous histio-

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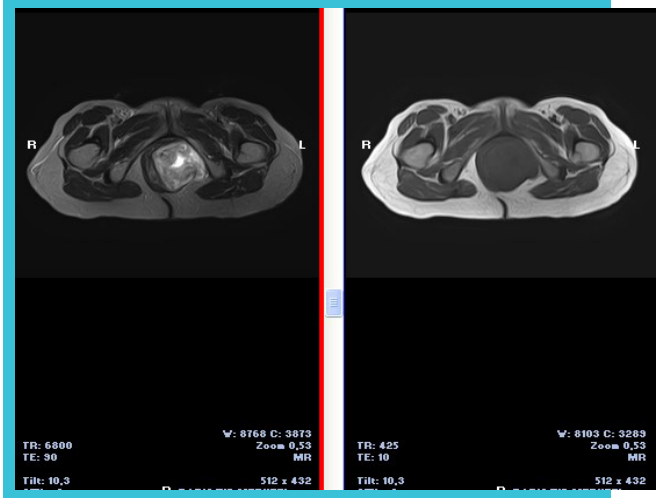
cytoma and other soft tissue tumors. In this paper a case of smooth muscle tumor of uncertain malignant potential of the vulva in a 33-years-old woman, which was misdiagnosed as an aggressive angiomyxoma was presented.

Figure 1.



A well-defined mass-like lesion about 7x9 cm in size is seen left vulvovaginal wall. It shows bright signal intensity on T2-weighted image, and demonstrates delayed enhancement after contrast administration.

Case Presentation

A 33-year-old multipar woman with 2 children, presented with 8x9 cm mass on her left vulvo-vaginal wall, extending into retroperitoneal space. Abdominal ultrasonography revealed a 5 cm pelvic mass resembling lipoma. On pelvic magnetic resonance imaging, there was a 7x9 cm lesion between posterolateral bladder wall and ischioanal fossa (Figure 1). Biopsy was taken from the lesion revealing aggressive angiomyxoma. With the diagnosis of aggressive angiomyxoma the operation was performed, wide local excision of the mass was done extending posteriorly to the ischioanal fossa, superiorly up to clitoris and left vesical space, medially vaginal wall and laterally up to the ischial tuberosity (Figure 2). Complete resection was done and surgical margins were macroscopically found to be negative. Her preoperative and postoperative hemoglobin levels were 11.2 and 10.3 g/dl and patient didn't need any transfusion. The pathology result was reported as STUMP with tumor free margins (Figure 3). No adjuvant therapy was administered and the patient made an uneventful recovery, with no recurrence so far for 12 months.

Figure 2.

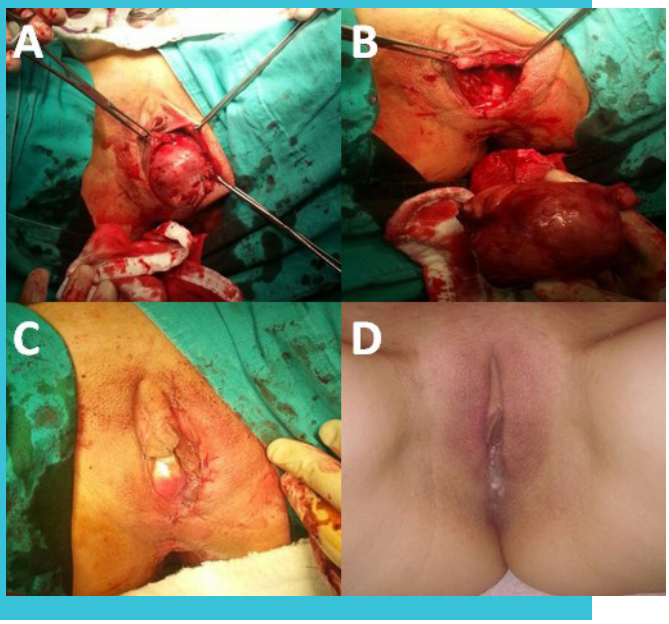


Figure 2A-2B: *Excision of the tumor, intraoperative view;* **2C:** *Post-operative view;* **2D:** *Postoperative view at 6 months.*

Discussion

The smooth muscle tumors of uncertain malignant potential tumors are rare lesions of the vulva. Most tumors are small (<5 cm), firm and well circumscribed. Clinically aggressive tumors tend to be larger, more infiltrative and may show hemorrhage or necrosis [6]. The best accepted criteria for a diagnosis of uterine STUMP are defined by the Stanford criteria [3,7]. Three major factors are defined including cytological atypia, mitotic index and necrosis but these criteria are different for vulvar tumors. Nielsen proposed that the most common finding in smooth muscle tumors 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 HPF and grade two to three atypia [8]. Microscopically, most tumors are composed of elongated cells arranged in fascicles (conventional type), although an epithelioid component may be present. Myxoid or hyaline stroma is often present (myxohyaline pattern) and this may be especially prominent during pregnancy [9].

Figure 3.

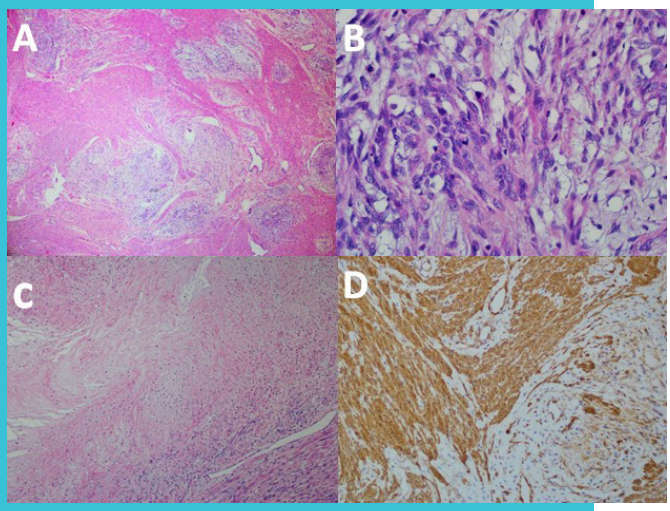


Figure 3A*: Epithelioid STUMP, hematoxylin eosin (H&E), X40;* **3B:** *Focal atypia and mitosis x400;* **3C:** *Focal necrosis H&E, x40;* **3D:** *Smooth muscle actin positivity (SMA), Immunohistochemistry (IHC), x200*

Almost all of these tumors express estrogen receptors or progesterone receptors. Because of the hydropic or myxoid change, especially during pregnancy, they may be confused with aggressive angiomyxoma on a small biopsy specimen. Imaging studies to assess the extent of the underlying lesion are often helpful in this differential diagnosis, because aggressive angiomyxoma tends to widely infiltrate deep soft tissue, whereas smooth muscle tumors form distinct masses [10,11]. Additional, deeper biopsies usually demonstrate the characteristic smooth muscle tumor. Most vulvar STUMP recur locally; less than one-fourth metastasize to the lungs or other distant sites causing death of the patient. However, long-term follow-up is limited in most series and the rate of metastasis may be higher than currently appreciated [12]. Although smooth muscle tumors with unusual histologic features may pose differential diagnostic problems, the major difficulty in diagnosing smooth muscle tumors of the vulva lies in predicting their recurrent potential with a reasonable degree of accuracy. Because the presence of any mitotic activity, nuclear atypia, necrosis, or evidence of infiltration into adjacent tissue may be associated with recurrence potential regardless of size, such lesions should probably be completely excised with clear margins whenever possible [13]. Due to the low prevalence of these tumors, there are no evidence based diagnostic algorithms or published recommendations for treatment. However, prior reports have recommended surgical excision with the potential addition of radiation therapy. Decisions are made based upon the individual case presentation and pathological evaluation. Conversations between pathologists and clinicians can provide guidance to ensure adequate surgical excisions are performed. Prior studies have shown that risk of recurrence is most closely related to inadequate resection of margins. Close monitoring of the patient is advised, as these entities have almost a 50% recurrence rate [14]. In conclusion, the clinicians and pathologists should be aware of this rare differential diagnosis of vulvar mass. Although usually benign, the tumor requires thorough pathological and radiological workup to rule out malignant features and other close differential diagnoses. Early aggressive resection is recommended due to destructive nature of this tumor on adjacent tissues. The tumor has a tendency for recurrence and hence the resected margins should be clear and long term follow-up should be arranged. Although the experience with follow-up of this tumor is scarce, the prognosis depends on complete surgical resection and lack of pathological features of malignancy.

Acknowledgement
None

Conflict of Interest Statement
The authors declare no conflict of interest

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