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Case Report

Aggressive angiomyxoma in a male patient: A rare case

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Abstract Aggressive angiomyxoma (AAM) is a rare mesenchymal tumor commonly affecting perineum of female, but also inflicting analogue area of male in lower incidence. We describe a 22-year-old man with a pedunculated, soft tumor, sized 10x5x7 cm in largest part, on his gluteal sinistra area. The tumor was completely excised, and histology examination showed squamous epithelia, spindle- and stellate-shaped cells within myxoid stoma without mitotic figures, fibrosis in some area, and thick-walled blood vessels. No recurrence was observed until 1-year follow-up after surgery.

Key words
Aggressive angiomyxoma-male-pedunculated tumor.

Introduction

Aggressive angiomyxoma (AAM) is a rare mesenchymal tumor characterized by tumor on vulvovaginal, perineum or pelvis of female. This neoplasm is designated as aggressive due to infiltrative nature and frequent local recurrence.[1] Most of AAM occurs in women, with female to male ratio is 6.6:1.[2] In men, AAM affects analogue area including inguinal, scrotal, and perineum.[3] Estrogen and progesterone receptors (ER and PR, respectively) were found in AAM suggesting that AAM might be hormone-dependent.[4] Diagnosis of AAM is established by using characteristic clinical and histological features.

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Case report

A 22-year-old man presented to our dermatology outpatient clinic with chief complain a lump on his left buttock disturbing when he was sitting. Since 1 year previously, the lump was gradually getting larger without any accompanied patient complains of pain and itching. There was no own and family history of same disease and skin cancer. Patient did not have relevant sexual and environment exposure. The patient was otherwise healthy. On physical examination on the gluteal sinistra there was a pedunculated tumor, sized 10x5x7 cm in largest part, with a 2 cm-long stalk and soft in consistency **(Figure 1)**. There was no regional lymphadenopathy. This patient was referred to department of surgery to undergo an excision.

After excision, grossly the tumor was covered by normal-looking skin and rubbery in palpation. Longitudinally sectioning revealed brownish-white mass. Histologic examination using hematoxylin-eosin staining showed squamous epithelium, whereas in dermis there

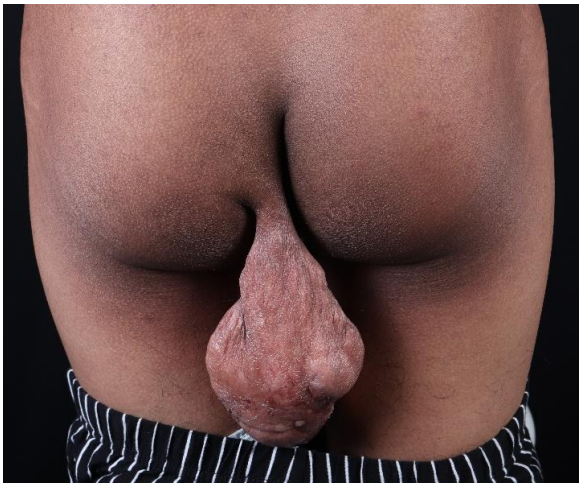

Figure 1 A pedunculated tumor in gluteal sinistra.

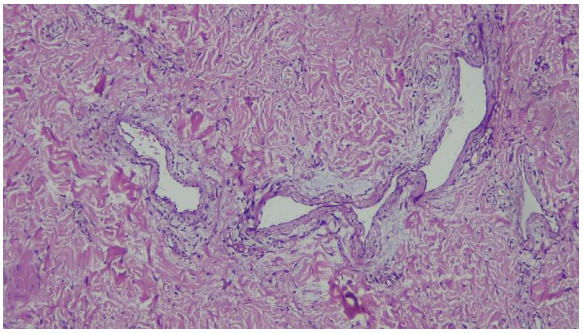

Figure 2 Histopathological examination in AAM.

were spindle- and stellate-shaped cells distributed sparsely throughout myxoid stroma, without mitotic figures, fibrosis in some area, and thick-walled blood vessels **(Figure 2)**. The features were suggestive of AAM.

Differential diagnosis of AAM in this case includes angiomyofibroblastoma, fibroepithelial stromal polyps, and myxoid lipomatous tumors. Patient condition after excision was good and no recurrence was observed until 1-year-follow-up after surgery.

Discussion

Aggressive angiomyxoma are locally invasive mesenchymal neoplasm which occurs commonly in female genital area. The term aggressive is derived from locally invasive nature and high recurrence of this tumor.[5] The cell of origin of AAM is still controversial. Initially the tumor is presumed to be myofibroblastic in origin.[6] Subsequently, fibroblastic and smooth muscle cell origin were proposed. Ultrastructural studies, however, suggest that AAM is derived from undifferentiated mesenchymal cell which differentiate into fibroblast or myofibroblast.[7]

The initial presentation of this tumor varies from an asymptomatic perineal or vulvar nodule/polyp or perineal hernia to a pelvic mass. Accordingly, diagnosis is established based on histological examination after surgical excision. Histologically, AAM presents with spindle- and stellate-shaped cells within myxoid background, prominent vascular proliferation, and devoid of mitoses.[8] Histopathologic features of AAM in men are similar to those of the conventional female cases. In our case, tumor presents as asymptomatic, pedunculated mass on left gluteal region, and after tumor reached a significant size, the patient found it disturbing when he was seated. Histology examination performed to the excised tumor revealed features which were appropriate with histologic characteristics of AAM.

We have to differentiate AAM from angiomyofibroblastoma (AMF), fibroepithelial stromal polyps (FSP), and superficial angiomyxoma (SAM). AMF is a benign, myxoid, soft tissue neoplasm of the vulva, which is well-circumscribed and usually smaller than 5 cm. AAM, on the other hand, usually has size larger than 5cm. Histologically, AMF presents with alternating hypocellular and hypercellular area of bland and spindle-shaped stromal cells with eosinophilic cytoplasm and minimal mitosis, surrounded thin-walled blood vessels.[9] Another important feature of AMF is multinucleated giant cell with linearly arranged nuclei.[10] These histologic features were not found in our case.

FSP is a relatively common benign, polypoid lesion of the vulvovaginal region. Histologic examination shows hyperplastic squamous epithelium and underlying edematous or myxoid stroma. The stroma consists of multinucleated stromal cells and variably sized blood vessels.[1] These characteristics differentiate fibroepithelial stromal polyps with our case.

SAM presents with a benign, solitary or multiple lesion which commonly occurs on the head, neck or trunk.[11] Histologically, the tumor is usually located in the dermis and subcutis and well encapsulated. The stroma is loose myxoid containing polymorphonuclear and mixed inflammatory cells. Within the stroma, stellate- and spindle-shaped cells are spread out and vasculature was not prominent.[5] Our case is not likely to be SAM because it is located clinically in gluteal region and histologically in deeper area, has different infiltrative pattern and prominent vasculature.

AAM is a rare tumor and commonly presents in female, however it can affect male in lower prevalence. Approximately 50 cases of AAM in men have been reported in the literature.[5] Clinically, AAM in men presents mostly in analogue area of female perineum and genital area, those are scrotum and inguinal region. Interestingly, AAM in men is mostly located in left side. This case is the first AAM in men that we found in our clinic and in our searchable hospital medical record. In our case, the tumor is located in the gluteal region, left side. AAM in female is likely to be hormone dependent as most of the tumor show estrogen and progesterone receptor positivity.[4] AAM in male has been shown to express estrogen, progesterone and/or androgen receptor and have immunohistochemical profile similar to those of female AAM.[5] We could not examine estrogen, progesterone and androgen receptors in our case, however as the tumor had been growing in pubertal period of the patient, we could assume that growth of the tumor might be influenced by sex hormones existing in the patient. The first line treatment of AAM is wide local excision with clear excision margin on histology. High rate of local recurrence generally occurs in the first year after primary surgery. Most recurrence is related to inadequate resection and residual tumor. Recently, however, a similar long-term prognosis was observed in patients with clear surgical and affected margins.[12] In our patient, surgical excision was performed by removing tumor and the surrounding healthy tissue near the stalk to achieve the R0 (microscopically margin-negative) resection. No recurrence was observed in our case in 1-year follow-up after surgery.

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