

Research Article**An Aggressive Angiomyxoma of Vulve; a Case Study****Azadeh Afzalzadeh¹, Mohammad Reza Bakhshandeh Rahimabadi^{2*},****Seyed_Saadat Gholami³ and Fateme Kiani³**¹Assistant professor of Obstetrics and gynecology,
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Yasuj University of Medical sciences, Yasuj, Iran*Corresponding Email: saadatg561@gmail.com**INTRODUCTION:**

Aggressive angiomyxoma is a benign tumor of the genital area with mesenchymal origin. The tumor often involves the vulve area, but other areas affected by it might involve perineum, buttock, and hips. Tumor mainly affects women of childbearing age and grows slowly and gradually (1).

Due to the high tendency to local invasion and recurrence after surgery, it is called aggressive (2). Its exact etiology remains unclear, but some pathologists have considered fibroblastic or myeloblastic as its sources (3).

Besides, sometimes the tumor responds to hormonal manipulation that is observed in estrogen and progesterone receptors in aggressive angiomyxoma (1,4). The main treatment of the tumor in case don't surgical contraindication is a full excision surgery.

Radiotherapy and chemotherapy may also be considered as adjuvant therapies, but are less recommended due to the low mitotic activity of tumor(3).

A case study

The patient was a 41-year-old multiparous woman that was admitted with complaints of masses of large genital area which gradually began to grow ten months ago. There was no history of pain and vaginal discharge, but the examinations revealed two polypoid and pedunculated lesions on major right labial area at the size of 4*4*7 and 5*3*5 cm with a soft consistency without pain. Its surface layer contained necrotic appearance along with the

injured areas with distinct borders. Vaginal and pelvic examination showed normal results. No inguinal lymphadenopathy was observed while tests and ultrasound of patient's abdomen were normal as well. The patient went under full masses excision surgery. Histopathology revealed the presence of thick walled vessels in different sizes in a stromal collagen and myxoid with spindle and starred cells and reported aggressive angiomyxoma. Surgical resection margin was free of disease. The reported patient has been asymptomatic and there is no evidence of recurrence from the time of the surgery to the date which is six months.

DISCUSSION

Steeper and Rosaie described aggressive angiomyxoma for the first time in 1983 (1). It is predominant in women of reproductive age, but some cases have also been reported in males (the ratio of female to male is reported to be 6 to 1). This asymptomatic tumor often grows gradually. Due to the high tendency to local invasion and recurrence after surgery, it is called aggressive. Tumor recurrence rate of 30% has been reported in studies (2). Histological presences of the tumor are diagnosed with the existence of spindle and starred cells in a loose and myxoidstroma. Tumor diagnosis is usually mistaken by Bartholin's, lipoma, and labial cyst. The vascular component of the tumor in histology façade helps distinguishing it from other soft tissue tumors such as Angiomyofibroblastoma, Fibrohistiocytoma, and myxolipoma that are other differential diagnosis of tumor (5). The tumor had distinctive features in various imaging and its

spread makes it easily determined. CT scan of aggressive angiomyxoma revealed that had well-defined margin with density less than of muscle and low signal on T1 MRI images. However, it had high signal on T2 MRI images which justifies the existence of soft myxoid matrix and high water content of its tumor. A hypo echoic mass is observed on ultrasound and angiography reports a mass hyper vascular (6). Some studies reported that the use of agonists GnRH drug therapy affects recurrence of the disease. Cases of metastatic tumors have also been reported (7,9,10). From the immunohistochemistry point of view, the tumor consisted of various combinations of estrogen and progesterone receptors, desmin, vimentin, and actin cd34 and cd44 (4&8). The exact pathogenesis is unclear, but a translocation on chromosome 12 with aberrant expression of a sequence of stirring isoform protein group of (HMGIC) I-C is observed in DNA transcriptions of the vulve area. A useful finding for the investigation of the remaining microscopic disease is the identification of the aberrant expression of HMG-C protein using technique immunoperoxidase (9). Complete resection of the tumor-free margins is essential for a successful surgery. The need for adjuvant treatments including arterial embolization and hormone therapy is reported in some patients. Radiation therapy and chemotherapy have shown little impact due to low mitotic activity of tumor, but is recommended for patients resistant to hormonal therapy and embolization. Long-term patient follow-up with MRI is useful for recurrence detection (3).

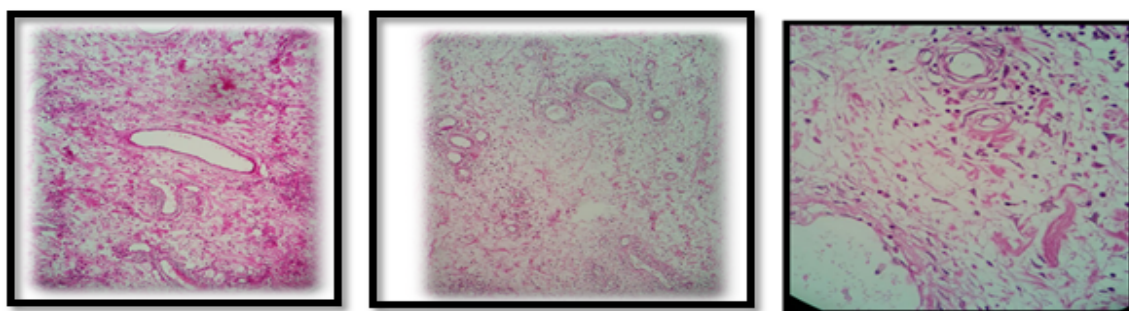


Figure 1,2,3: H & E pictures of angiomyxoma: ill defined paucicellular neoplasm composed of uniform appearance spindle cells with ovoid nuclei and prominent cytoplasm in hypocellular myxoid matrix. Also myxoidstroma contain variably size blood vessels.

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