

CASE REPORT

AGGRESSIVE ANGIOMYXOMA OF THE UTERUS - A RARE CASE

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ABSTRACT: Aggressive Angiomyxoma is a rare myxoid mesenchymal tumor of the pelvis and perineum, which occurs almost exclusively in adult women. It is a rare neoplasm with about 250 reported cases. To the best of our knowledge only one case of aggressive angiomyxoma of uterus arising from the uterine cavity and presenting as endometrial polyp has been reported. But angiomyxoma arising from body of uterus has never been reported. We report a case of 45 year old lady with a huge mass arising from the uterus which on histopathological examination showed features of aggressive angiomyxoma. Complete excision is the treatment of choice because of its tendency to recur locally. So radical surgery, with wide margins is advised. Prognosis is very good. Only 2 cases with metastatic disease followed by death have been reported. Recurrence rate is 36-70%.

INTRODUCTION: Aggressive Angiomyxoma (AA) was first reported in 1983 by Steeper and Rosai^[1]. It is a rare mesenchymal tumor of the pelvis and perineum that occurs almost exclusively in adult women². The less common sites are vagina, urinary bladder, perianal region in men particularly the scrotum.³ The adjective 'Aggressive' emphasizes the neoplastic character of the blood vessels, its local infiltrative nature and the high risk of local recurrence, not indicating the malignant potential of the lesion^[2]. To the best of our knowledge only one case of AA of uterus arising from the uterine cavity and presenting as endometrial polyp has been reported ^[3]. But AA arising from body of uterus has never been reported.

CASE REPORT: A 45 year old, parous woman presented with a painless swelling in the abdomen since 1 month. On clinical examination a soft mass arising from pelvis which was about 24 weeks pregnant uterus in size, non tender with variable consistency and with restricted mobility was seen. Speculum examination showed cervix pulled up. On per vaginal examination, same mass was felt in

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the posterior fornix, uterus could not be made out separately from mass. Provisional diagnosis of a malignant ovarian tumor or a fibroid uterus was made. Ultrasonography showed a large solid ovarian mass having heterogeneous echo texture measuring about 20x15 cm, originating from the pelvis and extending into the abdomen. Uterus was displaced towards left side and anteriorly. The impression was a large solid ovarian tumor/ carcinoma ovary.

At laparotomy, uterus of 6 weeks size with upward displacement was seen. Adnexa were normal. A fixed encapsulated tumor of size 20x20x11 cm with bosselated surface with congested vessels was seen attached to the posterior wall of the uterus, occupying whole of lower abdomen and upper vagina. No metastasis or lymphadenopathy was seen. Mass was separated from the posterior wall of uterus and simultaneously the steps of total abdominal hysterectomy with bilateral salpingo-oophorectomy were continued and mass was removed en mass.

Cut section revealed pale to white to brown solid gelatinous appearance with areas of hemorrhage and micro cysts filled with pale myxoid material. Microscopic examination showed partially encapsulated tumor showing predominantly moderate cellularity. Tumor cells were uniform, small, and stellate and spindle shaped having indistinct border. Nuclear atypia, binucleation and multinucleation not seen. Cells were seen scattered singly and in small clusters in abundant angiomyxoid and fibromyxoid stroma. Vessels of varying caliber showing perivascular hyalinization and vascular hypertrophy were noted in the stroma. Congested blood vessels and extravasations of red blood cells were seen. Necrosis and mitosis were not seen. The features were of an aggressive Angiomyxoma.

The patient had an uneventful post operative period and went home 9 days later.

DISCUSSION: AA is a rare neoplasm with about 250 reported cases [4]. It is most often found in women in reproductive age with a peak incidence in the fourth decade of life [5]. The female: male ratio is 6:1 [6]. In the latest WHO classification AA is classified under 'tumors of uncertain differentiation'.

The majority of patients present with a slow-growing mass which is otherwise asymptomatic and this is frequently the only symptom/sign. Observed accompanying symptoms and signs are regional pain, a feeling of local pressure, or dyspareunia [7]. It is worth noticing that the frequency of symptoms and signs attributable to local growth is lower than what would be expected from the relatively large size of most of these tumours [7].

Most AA are big, often more than 10 cm in largest diameter. These tumors are macroscopically lobulated and may adhere to surrounding soft tissue [7]. Microscopically, cells with a spindled or stellate morphology are seen, embedded in a loose matrix consisting of wavy collagen and edema. Cellularity is generally low to moderate. Infiltration into fat, muscle, and nerves is seen. The hallmark of AA is vessels of varying caliber haphazardly scattered throughout the tumor parenchyma, whereas mitotic figures are scarce [7]. The diagnosis is based on histopathological and immunohistochemical studies [8]. Although immunohistochemical studies have revealed that tumor cells are immunoreactive for no specific marker, it can show positivity for desmin, vimentin and smooth muscle actin, CD 34, estrogen and progesterone receptors.

Although the exact etiology is not known, clonal cytogenetic aberrations have been reported in some aggressive Angiomyxoma, including loss of an X chromosome, a translocation involving 12q14-15 [10], rearrangement of the HMGIC gene [2]. Using immunohistochemistry or ultra structural

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analysis, it was proposed that the neoplastic cells of AAM exhibit fibroblastic and myofibroblastic features and appear to be hormonally influenced [10]. The possibility that a progenitor cell has the capacity for smooth muscle differentiation has also been raised [8] favoring isolated primitive mesenchymal cell origin of the tumour.

Complete excision is the treatment of choice because of its tendency to recur locally [6]. So radical surgery with wide margins is advised [14]. Because most tumors are large, grow infiltrative and blends with adjacent soft tissue, and are located in close proximity to vital organs such as bladder and rectum, wide excision is not always possible and/or may cause significant morbidity[7]. In such situations watchful waiting may be advisable because these tumors may be stable with no or very limited growth over long periods of time [7] or incomplete or partial resection can be tried[6]. Several reported attempts using chemotherapy and radiotherapy as part of the treatment for AA have been disappointing, probably due to the low mitotic activity/growth fraction of cells[7]. Most AA express estrogen and progesterone receptors and are likely to have a hormone-dependent growth [6] because of this, treatment with GnRH agonists has been tried to prevent and also treat recurrent AA.

Prognosis is very good. Only 2 cases with metastatic disease followed by death have been reported [7]. Recurrences are common, reported to be between 36-70%[6]. There is no correlation between size of tumor and recurrence rate [6]. Late recurrences may develop several years after the primary tumor was found, and long-term follow-up of the patients is therefore very important. 1-2 yearly intervals of follow-up with alternate MRI and sonography is sufficient in view of the slow growing and non metastasizing nature of this tumor.[11]

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Fig. 1: Angiomyxoma presenting as mass per Abdomen

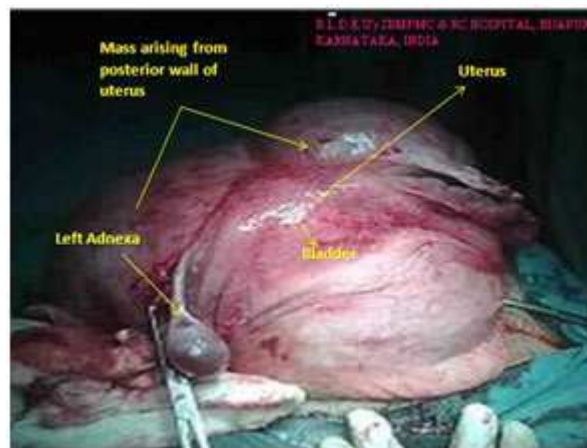


Fig.2: Intra-operative view of the mass

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Fig. 3: Posterior view of the mass

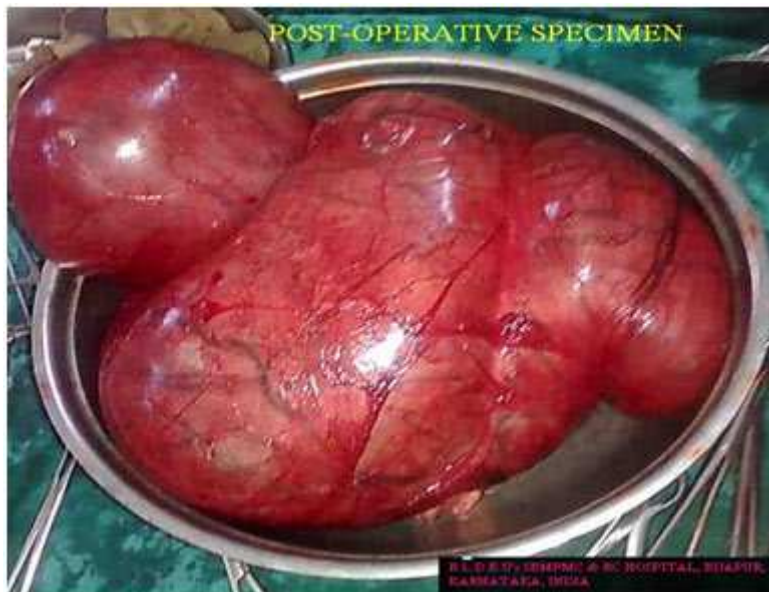


Fig. 4: Specimen of angiomyxoma

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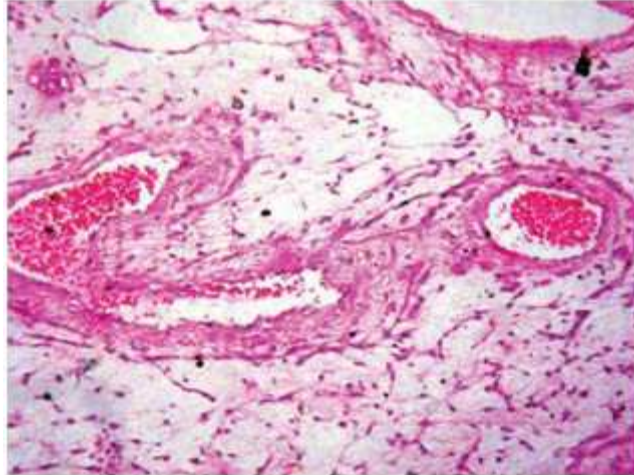


Fig. 5: Photomicrograph showing paucicellular tumor with myxoid matrix

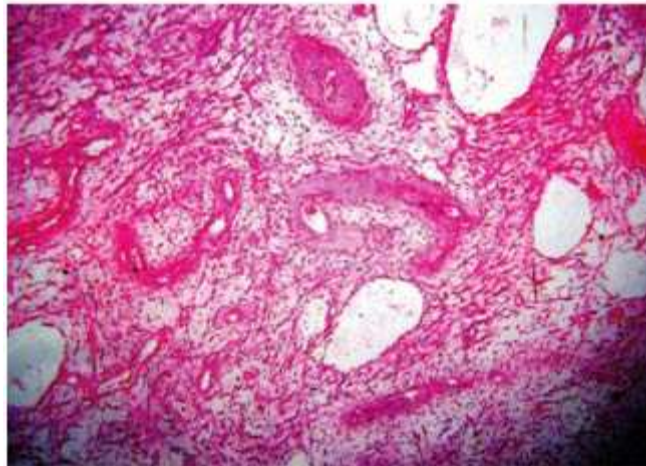


Fig.6: Photomicrograph showing vessels of varying caliber & perivascular hyalinization