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An Overview of Aggressive Angiomyxoma (AAM)

KEYWORDS: Angiomyxoma, abdominal lump.

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ABSTRACT *Aggressive angiomyxoma (AAM) is a benign soft tissue neoplasm. In latest WHO classification AAM has been classified under 'Tumours of uncertain differentiation'. Other name for this entity is 'Deep aggressive angiomyxoma'. Only around 192 cases have so far been reported from all over the world. The term "aggressive" has been labelled to this neoplasm because it has very high probability of local recurrence. Metastasis is very uncommon. The tumour mainly arises from female pelvis and genital area. It has been also been reported to arise from extrapelvic & extragenital areas. The tumours are usually huge in size, more than 10 cm in diameter, and push adjacent organs rather than invading those. Microscopic features include vascular component (vessels of varying diameters) and cellular component in huge myxomatous stroma. Diagnosis is helped by ultrasound and MRI. Certain immunohistochemical and features are characteristic. Treatment is wide excision. Close follow-up over several years is essential because of risk of recurrence.*

INTRODUCTION

Aggressive angiomyxoma (AAM) is a benign soft tissue neoplasm. In latest WHO classification AAM has been classified under 'Tumours of uncertain differentiation'. Other name for this entity is 'Deep aggressive angiomyxoma'. (1)

The term aggressive was introduced to emphasize the locally aggressive behaviour and high potential for local recurrence. However, it does not infiltrate surrounding organs. It has a very low probability of metastasis. Out of 192 cases reported so far, including our one case, only 2 cases with metastasis have been reported. (2) The name angiomyxoma was chosen as these are similar to myxoma with a notable vascular component. (3)

ETIOLOGY

No etiologic factors, as far as present knowledge goes, are known.

EPIDEMIOLOGY

AAM is uncommon neoplasm with about 192 cases reported.

Hussamuddin Adwan et al, from Department of Surgery, St Mary's Hospital, London, UK have reported 180 cases of aggressive angiomyxoma after web search.(4) Including first report by Steeper and Rosai in 1983.(5) Of these 70 are pelvic or retroperitoneal in the year 2004. Since then 12 cases have been added by Amezcua et al.(6) Of these 2 were vulvar, 1 suburethral, 2 perirectal. So 75 cases have been reported since the first 9 cases reported by Steeper & Rosai in 1983 , we have added the 76th case which occurred in genital area. It is important to note that we disregarded cases arising from extragenital area. If we include all types, the total number of cases becomes 180+12=192.

The pelvic AAM is usually found in women in reproductive age with peak incidence in the fourth decade of life. The female to male ratio is 6:1.

CLINICAL FEATURES

These may present as lump anywhere in pelvis, perineum, vulva & vagina. These tumours are slow growing & painless. Many times a Bartholin cyst is taken for marsupialization and is found to be some other thing and then retrospectively investigated & turns out to be AAM. Abdominal lumps are mistaken as ovarian tumours and at laparotomy it is found that the lump is preperitonial as it occurred in our case. Many cases are reported to have occurred after Total hysterectomy with bilateral salpingo-ophoectomy.

Most AAM are large tumours often attaining size of 10cms or more as they rarely cause symptoms.

Sign & symptoms include pain (although rare), feeling of local pressure or dyspareunia.

Recurrences should be followed up 6 monthly radiologically to avoid these being missed.

Differential diagnosis:

1. Myxoid tumor
2. Angioblastoma

RADIOLOGY

USG reveals large well-defined mixed echogenic lesion.

MRI- findings show isointense or hypointense lesion on T1-weighted MRI. These tumours show contrast enhancement reflecting their inherent vasculature. The tumours tend to displace and grow around structures rather than infiltrate them.

CYTOGENTICS

Six cases of AAM having non-random involvement of chromosomal band 12q15has been seen. Other aberrations seen are t(11;12)(q23;q15),t(7;12)(q22;q13-14,t(8;12)(p12;q15) and der(12)t(5;12)(q31;p11)inv(12)(p11q14).

PATHOLOGY

Gross- These are generally huge tumours measuring more than 10cms in diameter. They are lobulated and may adhere to surrounding soft tissue.

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Microscopic features- The hallmark of these tumours is blood vessels of varying size scattered in myxomatous parenchyma. The mass is generally hypocellular. The cells with spindled or stellate morphology are seen in loose myxoid stroma.

Usually there is no evidence of nuclear atypia or mitosis.

IMMUNOCHEMISTRY- Immunohistochemical studies show strong staining for desmin, estrogen receptors, progesterone receptors, muscle-specific actin and smooth muscle actin. Some tumours are positive forCD34. Staining for S-100 protein is negative. These findings suggest myofibroblastic differentiation of these neoplastic cells.

TREATMENT

Radical surgery with wide margins is the treatment of choice. Because of close proximity of these tumours to adjacent organs such as bladder and rectum, at times wide excision is not possible. In such scenario close follow-up is warranted. Radiotherapy and chemotherapy have no role in management. Many tumours express estrogen and progesterone receptors and may respond to GnRH analogues.

PROGNOSIS

Prognosis is good. Only 2 cases with metastatic disease followed by death have been reported. Recurrence in 6 months to several years has been reported in 9 to 72% of cases. Hence, close follow-up is very essential.

IMAGES:

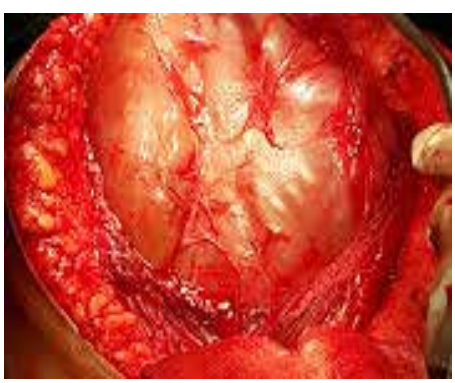

Fig 1. Lump on incision

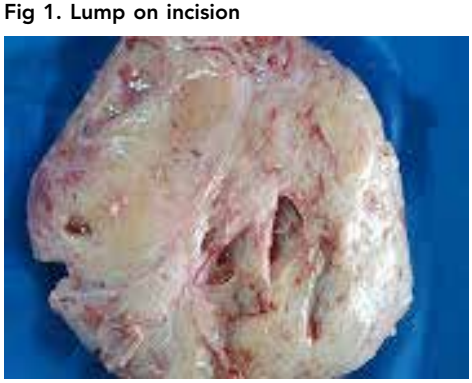

Fig 2. Cut section of the tumour

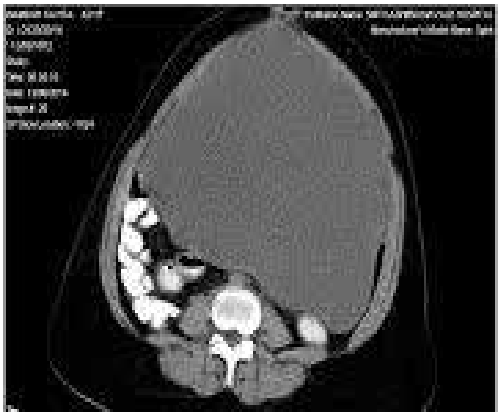

Fig 4. CT picture

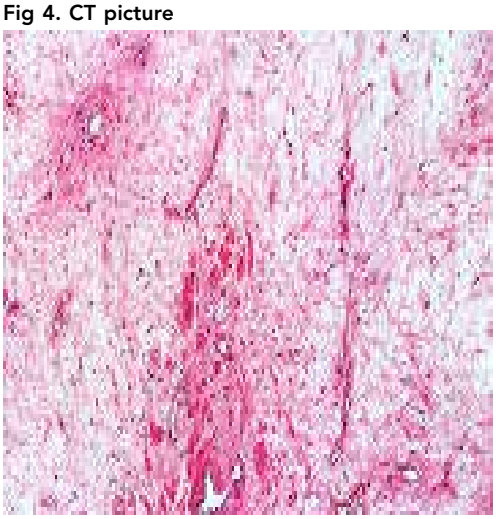

Fig 5. Microscopy picture 1

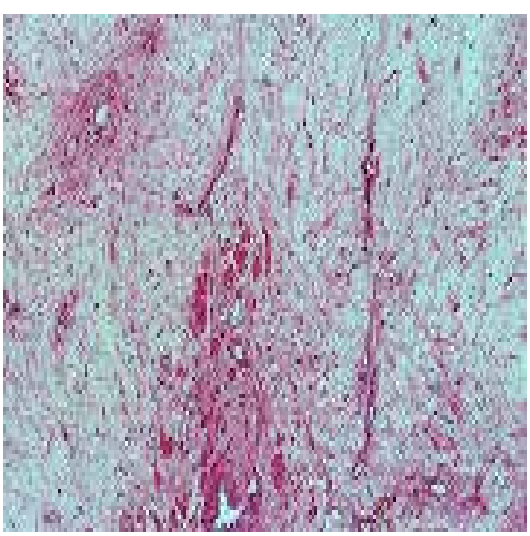

Fig 6. Microscopy picture 2

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