Aggressive Angiomyxoma in Men


Abstract
Aggressive angiomyxoma is an uncommon neoplasm that preferentially involves the pelvic and perianal regions of reproductive age females. These tumors are rare in men and merit a wider recognition in male urologic pathology. We report a case of a 65 year old male who presented with a swelling in the left perineal region since 2 years which gradually increased in size to 10x10x8 cms. CT scan revealed a well defined lobulated heterogenous minimally enhancing mass in the left ischiorectal fossa extending upto the subcutaneous plane. Microscopically stellate to spindle shaped cells arranged in a loose myxoid stroma with numerous thin to thick walled vessels were seen. There was no evidence of pleomorphism, hyperchromatism or mitotic activity. This case is reported because of its rarity in males. It is a benign tumor and requires a wide excisional surgery.

INTRODUCTION
Aggressive angiomyxoma is a locally infiltrative mesenchymal tumor which is common in the perineal region in young females and is rare in males. Approximately 150 cases have been reported in women. To our knowledge 40 cases of AAM occurring in men have been reported in literature. Sites frequently involved are the scrotum, spermatic cord, inguinal region and perineum. Gross and microscopic features are similar to those described in females.1 Female to male ratio is 6:6.1. Age distribution is wide ranging from 18 to 63 years with peak incidence at 31 to 35.2 The diagnosis of this tumor can be done on histology alone without the support of ancillary studies. This case is reported because this entity is rare in males and has an individual standing as it has to be differentiated from benign myxoid tumors because of its high tendency to recur and requires a wide excision surgery. On the other hand it should be differentiated from malignant myxoid tumors because it lacks the metastatic potential and any adjuvant therapy is not useful.

CASE REPORT
A 65 year old male presented with a swelling in the left perineal region since 2 years. The swelling gradually increased to attain the present size of 10x10x8 cms. There was no history of any bowel or bladder obstruction or any other systemic symptoms. On physical examination, skin over the swelling was freely moveable; there was no ulceration, bleeding or pain. CT scan revealed a well defined lobulated heterogenous minimally enhancing mass in the left ischiorectal fossa extending upto the subcutaneous plane and displacing the anal canal; however the fat planes between the mass and anal canal were well maintained. There was no evidence of any calcification.

We received specimen of a large mass measuring 13x10x9 cm. The mass was unencapsulated, externally lobulated, grayish white and glistening. Cut section was gelatinous (Figs. 1 and 2).

Microscopic examination revealed a low grade spindle cell lesion comprising of areas of hypocellularity amidst myxoid change and interspersed by numerous thin to thick walled vessels. There was no evidence of pleomorphism, hyperchromatism or mitotic activity. The periphery of the tumor showed infiltration into the surrounding fat (Figs. 3 and 4).

On immunohistochemistry, the cells were focally positive for smooth muscle actin and negative for S-100.

DISCUSSION
In 1983, Steeper and Rosai described a rare but distinctive, locally infiltrative mesenchymal neoplasm which had predilection for the pelvis and perineum in young females.3 Approximately two years later Begin et al published their experience with 9 examples, including the first reports of this entity in males.4 The other sites of involvement for this tumor are vulva, vagina, inguinal area, buttock, and peritoneum. As the tumors are often large, contiguous involvement of several sites is common.
The present case of AAM in a male arising in the left perineal region is characteristic of the tumor arising in the pelvic soft tissues originally described by Steeper and Rosai and later by Begin et al.

The female patients present with long standing chronic mass, pain in perineal, labial or pelvic region, pressure like and pulsating sensations, dyspareunia and increased mass effect with heavy lifting, urinary frequency, etc.

Commonly these tumors are more than 10 cms in size. These tumors have an overall lobulated architecture with relatively sharp margination in some areas and adherence to, or infiltration of fat, fibrous tissue and or muscle in others. A few tumors are reported to have diffusely infiltrative margins.4

Microscopically, these tumors have low to moderate cellularity. The neoplastic cells usually exhibit spindled or stellate morphology with relatively scant eosinophilic cytoplasm in a myxoid background. Large and medium sized blood vessels with a variety of secondary changes including medial hypertrophy and fibrointimal proliferation are scattered throughout the tumor. Mitosis may be present. Small scattered bundles or fascicles of smooth muscles may be evident. The evidence of infiltration can be seen by the entrapment of fat, muscle and or medium sized nerve bundles.

The histogenesis and degree of differentiation which may be exhibited in aggressive angiomyxoma have been debated in literature but a derivation from stromal cells possibly unique to the region favoured.

In males scrotum, spermatic cord, inguinal region and perineum are the sites frequently involved with case reports in retrovesical region, prostate and epididymis.1,5,6 Gross appearance and histomorphologic features are similar to those described in females with a male to female ratio of 6.6:1. Age distribution is wide ranging from 18 to 63 years with a peak incidence at 31 to 35.

One must rule out malignant lesions like myxoid liposarcoma and myxofibrosarcoma which are close mimicks. The vascular pattern of myxoid liposarcoma which has thin walled capillaries with small arcs and that of myxofibrosarcoma which shows less frequent thick walled with wider arc capillaries, helps differentiating it from aggressive angiomyxoma where the blood vessels are numerous, open and more rounded. Angiomyofibroblastoma which has similar appearance and site predilection can be ruled out on the basis of higher cellularity, abundance of vessels, plump stromal cells and absence of extravasation of RBC’s.7

Immunohistochemistry shows positivity for desmin, vimentin, smooth muscle actin, muscle specific antigen,
estrogen receptor, and progesterone receptor and negativity for S-100, Ki-67(MIB). Hormone reactivity is important and suggests that AAM may arise from specialized hormonally responsive stromal cells of the perineum and may play a therapeutic role in unresectable tumors.1,8

In our case, the diagnosis was based on the microscopic findings: low grade spindle cell lesion comprising of areas of hypocellularity amidst myxoid change, interspersed by numerous rounded thick and thin walled vessels; and immunohistochemistry has supported the diagnosis.

In conclusion, this case is reported because this entity has an individual standing as it is rarely seen in males with very few cases reported in literature and has to be differentiated from benign myxoid tumors because of its high tendency to recur when incompletely excised. (recurrence rate 36 to 72%). On the other hand it should be differentiated from malignant myxoid tumors because it lacks the metastatic potential and any adjunctive therapy is not useful. With adequate sampling and sufficient clinical information, a histologic diagnosis of aggressive angiomyxoma can usually be made without the need of ancillary studies. The role of hormonal therapy in the management of this peculiar tumor is yet to be determined.

REFERENCES


Announcement


For further details contact : Creative Travels Pvt. Ltd. Attn. Rama Satija, Creative Plaza, Nanakpura, Moti Bagh, New Delhi 110021, India.
Tel. : +91 11 2687 2257/58/59; Fax : +91 11 2688 5886; Email : apdw@apdw2008.net
Website : www.apdw2008.net

Announcement

Medicine Update 2008

Organized by Department of Medicine, All India Institute of Medical Sciences, New Delhi.

In association with Association of Physicians of India (API) Delhi State Chapter on 10th August 2008 (Sunday) 1 pm to 5.30 pm
Venue : JLN Auditorium, AIIMS, New Delhi.
Theme : Important Therapeutic Issues in Medicine
Special Workshop : Sleep Disorders

All are cordially invited

There is no registration fees for the main conference, however prior registration is mandatory
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Dr. (Prof.) SK Sharma, Organising Chairperson, Head, Department of Medicine, All India Institute of Medical Sciences.