Abstract: OBJECTIVE- Aggressive Angiomyxoma (AAM) is a rare mesenchymal benign myxoid tumour of the pelvis and perineum which occurs most commonly in adult females. Only about 180 cases were reported in medical literature. We report a middle aged female patient with recurrent perineal swelling whose histopathology was suggestive of aggressive angiomyxoma.

METHOD- Case report and review of medical literature. CASE REPORT- A 40 year old women patient presented with a recurrent swelling in the perineum. Examination showed a pedunculated swelling of size 5x4x3 cm protruding just lateral to the right labia majora, firm in consistency. Swelling was compressible, irreducible and there was no cough impulse. Cytology was inconclusive. MRI revealed 12 cm long and 2 cm thick sinus tract extending from right perineal region coursing right ischiorectal fossa and tracking between the vagina and rectum with extension of sinus tract into roof of right ischiorectal fossa. Tract showed hyperintensity in T2 weighted image. Planned for exploration and encapsulated tubular firm swelling was excised. HPE reported as Angiomyxoma. No local recurrence on 6 month followup. CONCLUSION- Aggressive Angiomyxoma (AAM) is a rare mesenchymal benign myxoid tumour of the pelvis and perineum which occurs most commonly in adult females with female : male ratio of 6:1.[1] They are large, slow growing with tendency for local recurrence. Only about 180 cases were reported worldwide.[1] We report a middle aged female patient with angiomyxoma who presented with recurrent perineal swelling.

INTRODUCTION

Aggressive Angiomyxoma is a rare mesenchymal benign myxoid tumour of the pelvis and perineum. Angiomyxoma was first described by Steeper and Rosai in 1983.[2] It occurs most commonly in adult females with female : male ratio of 6:1.[1] They are large, slow growing with tendency for local recurrence. Only about 180 cases were reported worldwide.[1] We report a middle aged female patient with angiomyxoma who presented with recurrent perineal swelling.

HISTORY

A 40 year female patient presented with a recurrent swelling in the perineum for the past 6 months. Patient had no history of fever, pain and trauma. There was no associated history of bowel and bladder disturbances. Menstrual history was normal. She had two normal vaginal deliveries. She was operated for the similar swelling 1 year back at the same site. Patient was not a known case of any chronic illness.

EXAMINATION

Examination showed a pedunculated swelling of size 5x4x3 cm protruding just lateral to the right labia majora, firm in consistency. Swelling was compressible, irreducible and there was no cough impulse. Cytology was inconclusive. MRI revealed 12 cm long and 2 cm thick sinus tract extending from right perineal region coursing right ischiorectal fossa and tracking between the vagina and rectum with extension of sinus tract into roof of right ischiorectal fossa. Tract showed hyperintensity in T2 weighted image.

Keyword: Aggressive angiomyxoma, perineal swelling, myxoid tumor
With the differential diagnosis of perineal hernia, ischio rectal abscess, soft tissue tumour and haemangioma we planned for exploration.

**MANAGEMENT**

With patient in lithotomy position, under aseptic precaution an elliptical incision was made around the swelling and found an encapsulated tubular firm soft tissue swelling extending from right side of perineum along the right lateral wall of vagina to the roof levator ani was noted with no evidence communication to pelvic cavity. Wide local excision of the mass was done upto the pelvic floor. After securing complete hemostasis wound closed in layers. Excised specimen sent for histopathological examination which reported as an unencapsulated neoplasm composed of scattered stellate to short spindle shaped cells admixed with plenty of thin walled congested blood vessels in background of myxoid stroma feature of Aggressive Angiomyxoma. Patient was on regular follow up and no local recurrence was noted on 6 month followup.

**DISCUSSION**

Angiomyxoma was first described by Steeper and Rosai in 1983.[2] Since then about 180 cases were reported in literature. It’s a rare benign soft tissue tumour occurs mostly in the genital and perineal area of female patients. Its incidence is about 6-folds higher in females than males.[1] Usually it occurs in female in the reproductive age group between 3rd – 4th decades. It most commonly occurs in retroperitoneum, bladder, vulva and vagina in female, and in testis, epididymis, scrotum and inguinal region in male. Most tumours are large (>10 cm) and grow slowly. Signs and symptoms at presentation includes a discomfort from the mass, a visible mass, or pressure effects on adjacent pelvic organs. They are slow growing and locally invasive hence have tendency for recurrence. There is no consensus on complete pathogenesis of this tumour. As this tumours are positive for ER and PR receptors they are thought to arise from a specialized mesenchymal cells of the perineal region or from multipotent progenitor cells which display myofibroblastic or fibroblastic features. This is supported by the fact that this tumour cells express desmin.[4] Clonal cytogenetics aberrations have been reported in some aggressive angiomyxomas, including loss of an X chromosome[16] and a translocation involving 12q14-15. Grossly they are well circumscribed non-lobulated and encapsulated/unencapsulated. On cross section they have a gelatinous appearance and greyish discoloration. Histologically they are hypocellular and highly vascular tumor with a **Myxoid stroma** containing cytologically bland stellate or spindled cells.[1] The tumor cells are characteristically positive for estrogen and progesterone receptors, desmin, muscle specific actin and vimentin and negative for S-100.[6] They show contrast enhancement in computer tomography that reflects their inherent vascularity. MRI is the investigation of choice. MRI will show contrast enhancement in computer tomography that reflects their specific actin and vimentin and negative for S-100.[6] They show stellate or spindled cells.[1] The tumor cells are characteristically vascular tumor with a discolaration. Histologically they are hypocellular and highly cross section they have a gelatinous appearance and greyish circumscribed non-lobulated and encapsulated/unencapsulated. On tomography its final diagnosis is usually on histopathological examination of excised specimen. Differential diagnosis of soft tissue tumours that occur in the perineal areas in an adult female patient would include angiomyxoma, angiofibroma, myxoid nerve sheath tumours, myxoid smooth muscle tumours, and angiolipoma.

Complete excision of the tumour is the treatment of choice. Though it is a benign tumour, recurrence rate of 30% has been reported if not completely excised. Regular follow up with 1-2 yearly interval MRI should suffice for recurrence as they are slow growing and no distant metastases. They also show good response to gonadotropin-releasing hormone agonist treatment which remarkably reduced the size of the tumour and recurrences.

**SUMMARY**

Aggressive angiomyxoma are rare cause of perineal soft tissue swelling especially in females. It should be kept in differential diagnosis and should be evaluated with an Ultrasonogram and MRI. Complete excision of the tumor is the treatment of choice.

**Bibliography**

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