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Case Report

An unusual presentation of a rare tumour: aggressive angiomyxoma

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ABSTRACT

Aggressive angiomyxoma is a very rare variety of soft tissue tumour exclusively involves the vulvovagina, perineum and pelvic region of women of reproductive age group. We report the case of a 48-year old woman presented with gluteal mass. Nature of the mass could not be diagnosed by clinical radiological and imaging evaluation, however it was found to extend from pelvis to gluteal region. It was removed surgically. Histopathology revealed the mass to be aggressive angiomyxoma, a rare variety of pelviperineal tumour extended from pelvis to the gluteal region. AAM presenting as a gluteal mass is extremely rare.

Keywords: Aggressive angiomyxomas, Pelvic soft tissue tumour

INTRODUCTION

Amongst various abdominopelvic and pelviperineal tumours aggressive angiomyxoma (AAM) is a very rare soft-tissue tumor that carries a high risk of local infiltration and recurrence. It almost exclusively involves the vulvovaginal, perineal and pelvic regions of women of reproductive age. It exhibits a slow insidious growth pattern with a locally aggressive course. There have been fewer than 250 cases reported in the world literature till date.[1]

CASE REPORT

A 48-year old woman, who had previous three normal deliveries, having no relevant medical and surgical history, presented with a painless gluteal swelling which progressively increased in size for the past 3- 4 years. She had no gynaecological complain with regular menstrual cycle. Neither she had any urological symptoms like dysuria and incontinence; nor bowel related symptoms. Physical examination revealed a soft mass of approximately 6x5 cms in the left gluteal region.

She was referred to this hospital with a diagnosis of gluteal abscess. However thorough clinical evaluation revealed the mass was non-tender without any local signs of inflammation. Per vaginal examination revealed the mass extending upward and could be felt at the left posterolateral vaginal wall. Upper pole of the mass could not be appreciated through vagina. Uterus found to be normal size and shape. She was further investigated to ascertain the nature and extent of the mass**.**

USG revealed a large gluteal mass extending to the pelvis. Further evaluation by MRI found a well-defined pelviperineal mass of 15.9 X 7.8 X 4.5 cm size. In the perineal region the mass lies in anal triangle in left ischeorectal fossa extending inferiorly through urogenital diaphragm and reaching into the soft tissue adjacent to natal cleft. Superiorly the mass is stretching and elevating the levator ani muscle. It is extending into the left hemipelvis until the level of the bladder dome. Pelvic component causing mild displacement of urinary bladder and vagina towards contralateral side. Fat planes were preserved (Figure 1).

With a provisional diagnosis of soft tissue tumour of pelvigluteal region, patient was taken up for surgery. On incising the skin and subcutaneous fat over protruding area of left gluteal region, a pale pink colour mass protrude out from the gluteal buldge (Figure 2). Mass was

held with a long artery forceps and pulled down gently with simultaneous finger dissection separating the mass from the surrounding tissue where it was loosely attached (Figure 3).

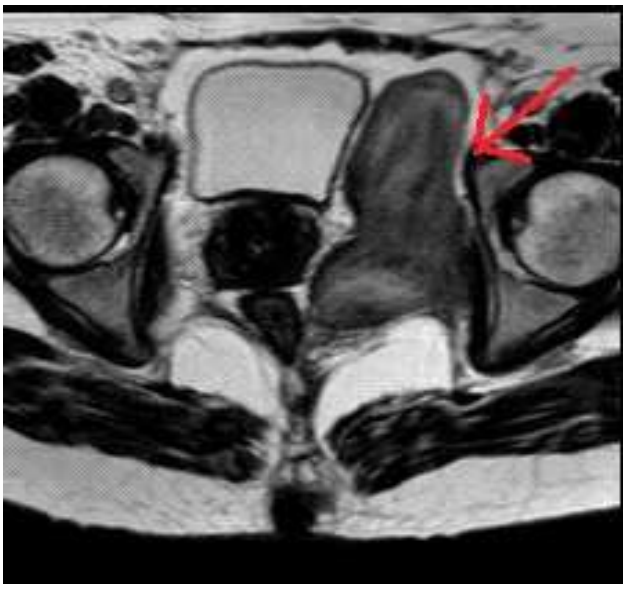

Figure 1: MRI showing a well-defined pelviperineal mass of 15.9 X 7.8 X 4.5 cm size.

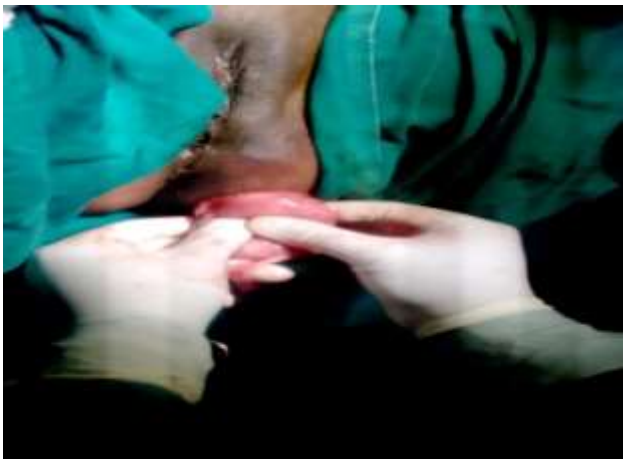

Figure 2: A pale pink colour mass protrude out from the gluteal buldge, on incising the skin and subcutaneous fat over protruding area of left gluteal region.

By gentle traction and blunt dissection the whole mass measuring 45 cm could be removed through the gluteal incision without opening the abdomen (Figure 4). No active bleeding was observed from the potential cavity left behind after removing the tumour. The dead space was packed with roller gauge. Skin incision was sutured keeping one end of the pack outside the incision, which was removed after 24 hours (Figure 5).

There was excessive serosanguinus discharge from the wound on first few days following surgery which dried up by 4 to 5th postoperative day. Stitches removed on 10th postoperative day. Review after one year showed no evidence of recurrence of the disease (Figure 6).

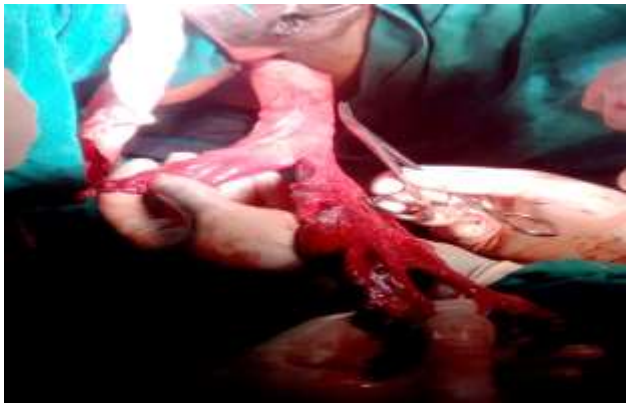

Figure 3: Mass was held with a long artery forceps and pulled down gently with simultaneous finger dissection separating the mass from the surrounding tissue where it was loosely attached.

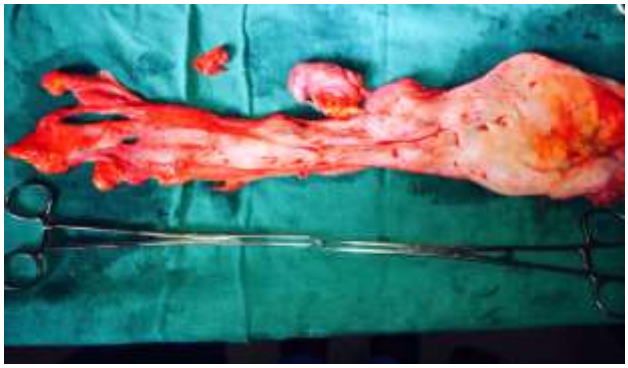

Figure 4: Whole mass measuring 45 cm could be removed through the gluteal incision.

DISCUSSION

Steeper and Rosai first reported nine cases of pelvic neoplasm, described as aggressive angiomyxoma in 1983.[2] The term is now widely accepted and, in 2003, was classified by the World Health Organization as deep angiomyxoma.[3] This tumour almost exclusively involves women of childbearing age (female to male ratio is 6.6:1). Very rare cases have been reported in perimenopausal women.[4]

These large size tumours are slowing growing and painless. The diameter of neoplasms has been reported as varying from 5 to 23 cm, but is usually >10 cm. The largest known AAM was described in 1998 as a mass measuring 57 × 47 × 23cm, which had increased slowly over approximately 8 years, most noticeably during pregnancy.[5] Commonest affected site involved are pelvis, perineum and vulva. Extension to the gluteal region has not been reported in the literature as is seen in our case. Although angiomyxomas are locally aggressive, distant

metastasis (to the lung) has been reported in only two cases.[6,7] It has a marked tendency to repeated local recurrence.

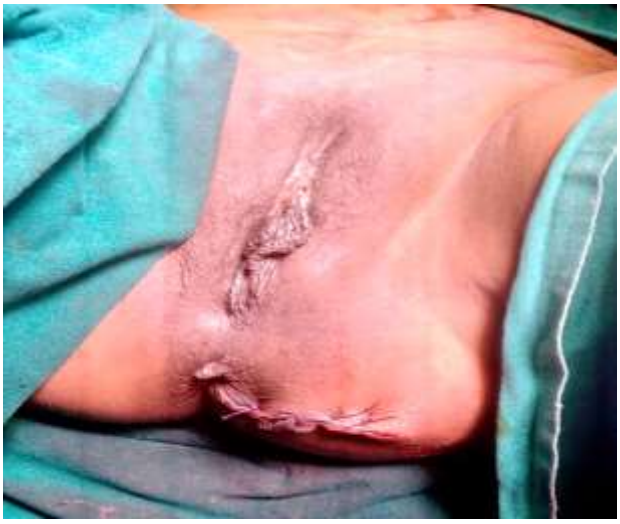

Figure 5: The dead space was packed with roller gauge. Skin incision was sutured keeping one end of the pack outside the incision.

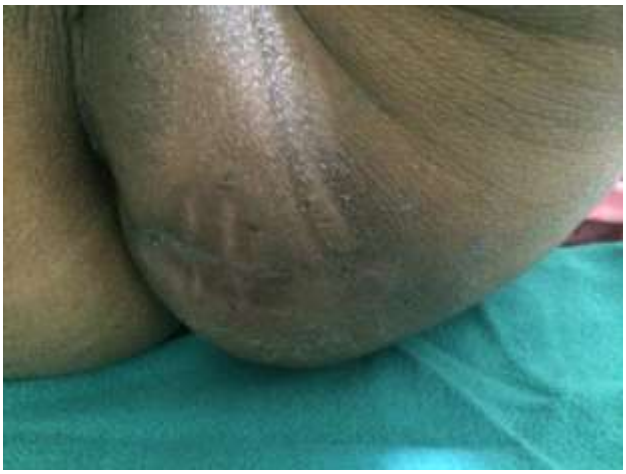

Figure 6: Review after one year showed no evidence of recurrence of the disease.

Macroscopically, AAM has a diffuse, gelatinous, homogeneous cut surface with areas of congestion and haemorrhage. Histological examination generally shows stellate and spindle-shaped neoplastic cells scattered in a background of loose myxoid stroma with numerous blood vessels of varying caliber (Figure 7). Mitoses and nuclear atypia are absent.

The stromal cells can show immunoreactivity to different combinations of Vimentin, Desmin, Smooth Muscle Actin, Muscle Specific Actin CD34, ER and PR.[8]

Preoperative diagnosis of AAM is often difficult because of its rarity. Misdiagnosis is common; occurring in more than 80% of cases.[8] The main differential diagnoses include vulvar abscess, lipoma, Gartner cyst, Bartholin cyst, vaginal cyst, vaginal prolapse, levator hernia and hernia of the canal of Nuck and other soft tissue tumour in this region like angiofibroblastoma, cellular angiofibroma, spindle cell lipoma etc.

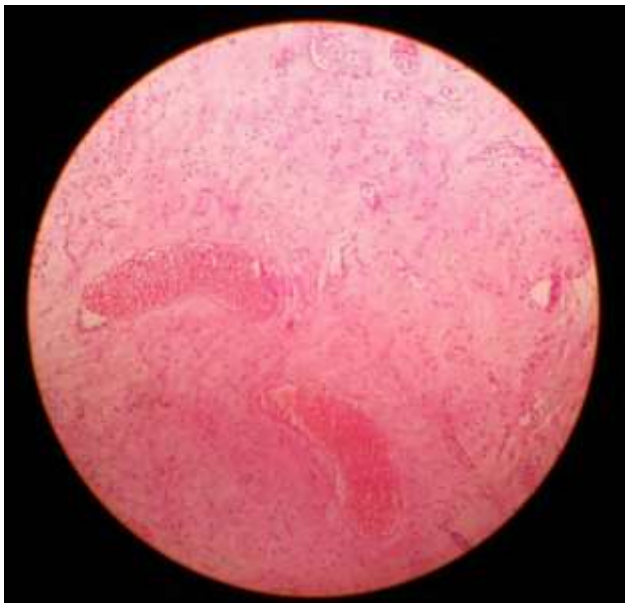

Figure 7: Histological picture loose myxoid stroma with numerous blood vessels of varying calibre.

Complete surgical excision is considered gold standard and remains the first-line of treatment. Radiotherapy and chemotherapy are unlikely to be useful adjuncts to primary surgery for AAM, because of low mitotic activity, but the use of radiotherapy to achieve local control or alleviate symptoms has been described. In view of the positive ER and PR status of some AAMs, primary treatment with GnRH agonists has been carried out successfully.[9] For example, McCluggage et al reported the case where after tumorectomy of the right vulva residual pelvic tumour was treated with 3.6 mg goselerin acetate once a month. Complete resolution on magnetic resonance imaging was achieved after 8 months of treatment, with no recurrence after 1 year. In addition, preoperative shrinking of tumors using GnRH agonists might increase the chance of complete excision and minimize the radical extent of the surgical procedure. There have been no previously published reports on the postoperative injection of GnRH for prevention of tumour recurrence.

The case described in the present report was treated surgically and no recurrence was observed till one year of follow up. Whether treatment is with surgery, hormone therapy or a combination of both, it is clear that AAM requires close, long-term follow-up to monitor for any recurrence and that the individualization of each case is essential for adequate management. This case is reported due to its rare presentation of an equally rare tumour of pelviperineal region.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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