

CASE REPORT

Aggressive Angiomyxoma of Rectovaginal Septum, Mimicking a Huge Recto-Enterocoele: A Rare Case Report

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Introduction

Aggressive angiomyxomas (AAM) is a rare mesenchymal tumor first described by Steeper and Rosai in 1983. Fewer than 150 cases have been reported in the world's medical literature [1–3]. They are mainly found in females in the pelvi-perineal region, usually in childbearing age [2]. It is a slow growing benign tumor with lack of metastatic potential. AAM is regarded as an aggressive neoplasm to denote the propensity for local recurrences. There is a strong female predominance with a female-to-male ratio of approximately 6:1. The rarity of the condition makes the preoperative diagnosis fairly difficult.

Case Report

A 41-year-old female presented with the complaint of something coming out vaginally for the past 11 years. It

was associated with difficulty in the urination and defecation. She is P2A1, without any significant history, general and per abdominal examination finding.

Per speculum examination revealed a huge swelling bulging out through the introitus from the posterior vaginal wall, reducible, no cystocele and no uterine descent, and uterus was anteverted, normal size on bimanual examination. With this, a clinical diagnosis of recto-enterocoele was made. All the routine investigations including chest X-ray were normal. Her abdomino-pelvic scan revealed a normal uterus, adenexa and ovaries, but a heterogeneous hypo-echoic mass of 126 × 77.2 mm posteriorly, thought to be a loaded colon (?). The rescan after bowel preparation reported to be normal.

A posterior repair was planned. While dissecting the posterior vaginal wall, a soft tumor mass was noted which was completely resected involving the surgeon. The pouch of douglas was high up without trace of enterocoele.

Grossly, the mass was globular, well-encapsulated measuring (15 × 13 × 9) cm and weighing 900 g. The cut section was homogenous glossy, with tan-to-white appearance and firm rubbery in consistency. Microscopic examination showed spindle cell with abundant myxoid stroma interspersed with medium-to-large-sized thick wall hyalinized blood vessels. Resection margin was free of tumor involvement. The histopathology report confirmed the tumor to be angiomyxoma of rectovaginal septum. Immunohistochemistry showed diffuse positivity for desmin and smooth muscle actin (SMA) positivity of the spindle cell surrounding blood vessels.

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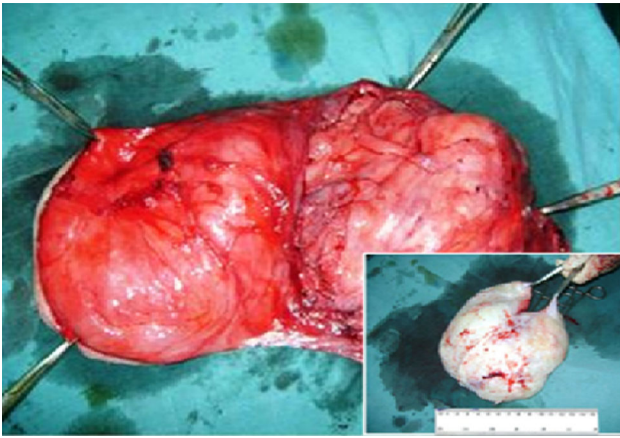


Fig. 1 Well-capsulated tumor and cut section showing homogenous glistening and solid appearance

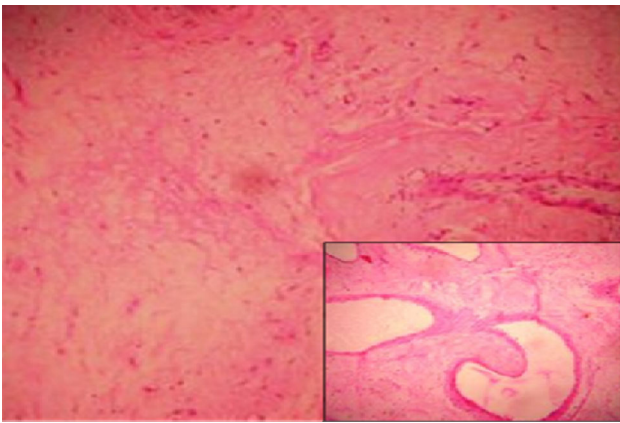
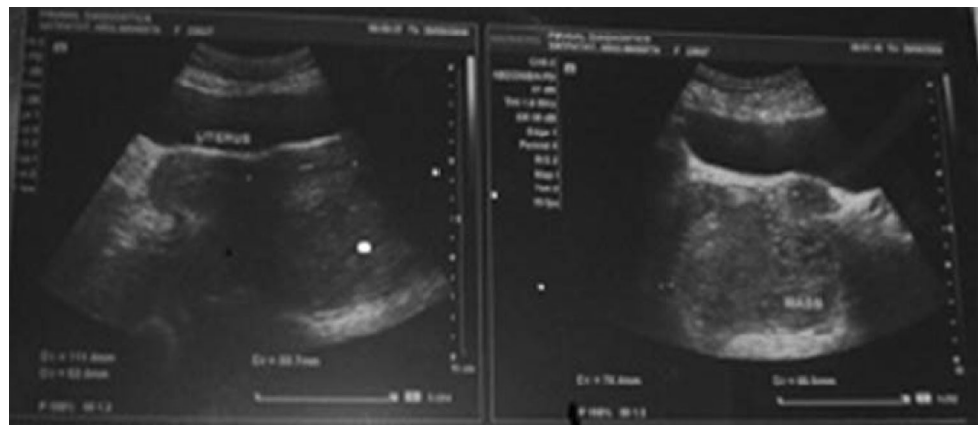


Fig. 2 Microphotograph showing myxoid stroma and few benign-looking spindle cells with highlighted area showing medium-to-large sized thick-walled vessels in a background of myxoid stroma (H&E stain)

Fig. 3 Ultrasound showing a mass posterior to uterus thought to be loaded colon



The patient had uneventful postoperative stay and followed up for last 2 years and was found to be asymptomatic without any evidence of local recurrence either clinically or radiographically (Figs. 1, 2, 3).

Discussion

AAM is a rare tumor found mainly in female pelvis. There are no large clinical trials to guide treatment, only case reports and small series, the largest of which identified 16 patients [3]. Surgical resection is the main stay of treatment [1–3]. Complete surgical excision with the tumor-free margin is the main objective [3], but local recurrence rates are high, varying from 35 to 72 % [1, 3], even with clear surgical margin. Resection of these cases can be technically difficult and challenging [1–3]. Value of extensive surgical resection to obtain clear margin has been questioned, as some cases with incomplete resection have not developed clinical recurrence over several years of follow up. Reports of metastatic disease further complicates the problem [3].

There are many unanswered questions about treatment and follow-up strategies. Long-term follow up with CT preferably MRI is recommended, since recurrences have been reported several years after first excision [1]. The patient we report has been doing well since last 2 years with no clinical and radiographic recurrence.

Conclusion

Although a rare diagnosis, aggressive angiomyxoma can present with unusual features. Histology is the gold standard for diagnosis. Wide excision is curative, and prognosis of such tumor is good.

References

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