International Journal of Reproduction, Contraception, Obstetrics and Gynecology
Revanna R et al. Int J Reprod Contracept Obstet Gynecol. 2022 Jul;11(7):2023-2026
www.ijrcog.org
pISSN 2320-1770 | eISSN 2320-1789

DOI: https://dx.doi.org/10.18203/2320-1770.ijrcog20221689

Case Report

Aggressive angiomyxoma of uterine corpus

Rachitha Revanna*, Padmasri Ramalingappa, Divya Tumkur Kumar

Department of Obstetrics and Gynaecology, Sapthagiri Institute of Medical Sciences and Research Centre, Bangalore, Karnataka, India

Received: 14 April 2022
Accepted: 04 May 2022

***Correspondence:**
Rachitha Revanna,
E-mail: rachitha26@gmail.com

ABSTRACT

Aggressive angiomyxoma (AA) is a rare slow growing mesenchymal tumour that preferentially involves the vulvovaginal, pelvic or perineal regions. It is called aggressive due its frequent local recurrences and infiltrative behavior. They occur most commonly in the reproductive age group in women. A 37 year old with AA of endometrial polyp, presenting with acute pain abdomen and friable, fleshy mass protruding out of introitus, precipitated by a dilatation and curettage for heavy menstrual bleeding. A clinical diagnosis of fibroid polyp with acute red degeneration was considered. There was a spontaneous expulsion of fleshy mass. HPE showed AA with features of torsion. MRI showed T2 heterogenous hyperintense mass lesion of 9 x 3 x 3.9 cm pedunculated polypoidal arising from upper anterior uterine wall. Hysterectomy specimen confirmed HPE findings of AA. Radiological studies and pathological evaluation aids in the diagnosis and planning of appropriate treatment of AA. Close and long term follow up of these patients should be emphasized due to high rate of local recurrence.

Keywords: Aggressive angiomyxoma, Mesenchymal tumor, Myxoid matrix

INTRODUCTION

AA is a rare slow growing mesenchymal tumour that preferentially involves the vulvovaginal, pelvic or perineal regions.[1] AA was first described by Steeper and Rosai in 1983. It is called aggressive due its frequent local recurrences and infiltrative behaviour.[2] They occur most commonly in the reproductive age group in women. Estrogen and progesterone receptors are commonly found in AA. It is thus likely to grow during pregnancy and respond to hormonal manipulation.[3]

Considering its locally aggressive nature, appropriate management and long-term follow-up is necessary. Many options for the treatment of recurrences have been tried with varying success, but no single modality is clearly beneficial over others.[4]

CASE REPORT

38 year old P2L2 came with complaints of severe lower abdominal pain for 10 days and a friable, fleshy mass protruding out of introitus, precipitated by a dilatation and curettage done 2 days back for heavy menstrual bleeding.

Local examination showed a well-circumscribed pedunculated polypoidal mass measuring about 5×6×4 cm with purple to blackish discoloration and bleeds on touch (Figure 1).

A clinical diagnosis of fibroid polyp with acute red degeneration was considered. There was a spontaneous expulsion of fleshy mass.

MRI showed T2 heterogeneously hyperintense pedunculated polypoidal mass lesion of 9×3×3.9 cm arising from upper anterior uterine wall (Figure 2).

HPE showed AA with features of torsion (Figure 3).

After expulsion of polypoidal mass, on per speculum examination, cervix and vagina were healthy, bleeding was present. On per vaginal examination, uterus size was corresponding to 14 weeks, anteverted, bilateral fornices were free and nontender.

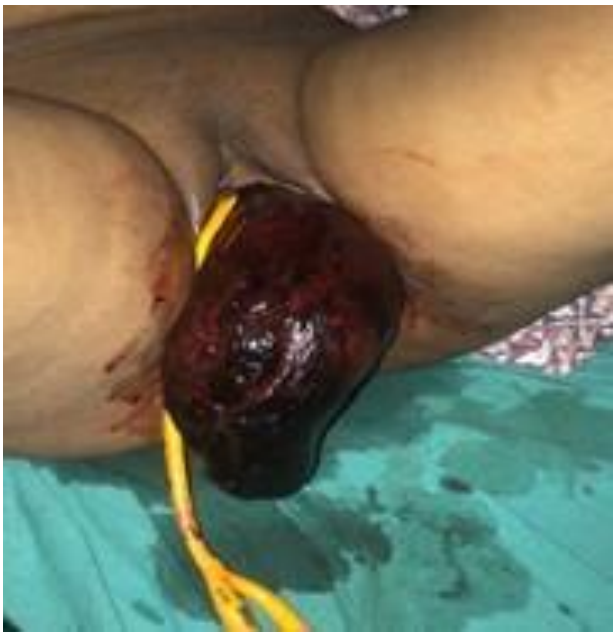

Figure 1: Clinical picture showing a well-defined pedunculated polypoidal mass measuring about 5×6×4 cm with purple to blackish discoloration present and bleeds on touch.

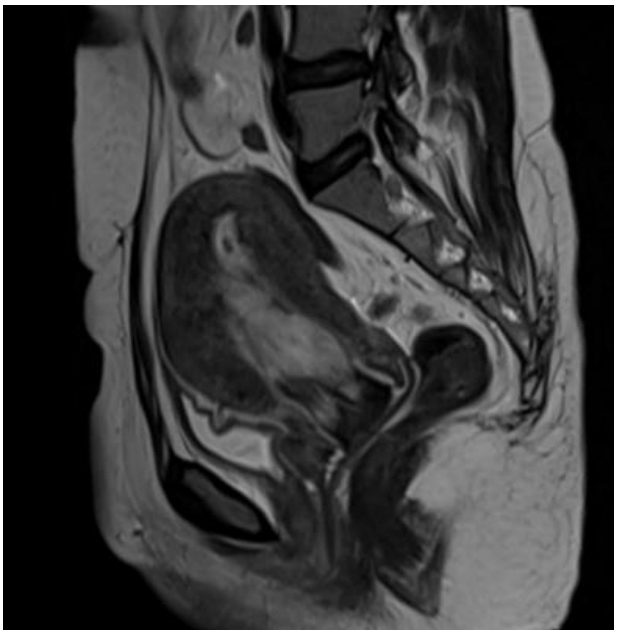

Figure 2: Sagittal T2w MRI showing heterogeneously hyperintense pedunculated polypoidal mass lesion of 9×3×3.9 cm arising from upper anterior uterine wall.

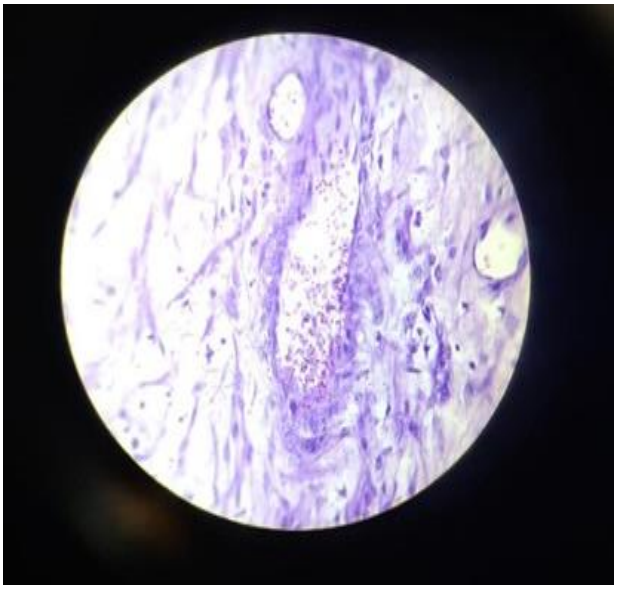

Figure 3: Microscopic picture stained with haematoxylin and eosin stain shows vascular channels dispersed in the myxoid stroma.

MRI pelvis was done after expulsion showed minimal residual mass in the uterine cavity. The patient was posted for laparotomy and proceeded with total abdominal hysterectomy and lymph node dissection.

Intra operatively, uterus size was 14 weeks with irregular surface and multiple small intramural fibroids were present. Bilateral external and right internal iliac lymph nodes were enlarged. Tubes and ovaries were normal. Omentum, bladder, bowel was normal.

DISCUSSION

Angiomyxomas are classified either as superficial or AA. Superficial angiomyxomas usually present in middle-aged adults as a single nodule or a polypoidal lesion in the head and neck region that may be clinically confused with skin tag or neurofibroma. The stroma is made up of mostly edema with little myxoid material. On the other hand, AA occured almost exclusively in the pelvic and perineal regions of women of reproductive age but was occasionally reported in men (male-to-female ratio 1:6).[5]

The term aggressive denoted its propensity for local aggression and recurrences after excision. Usually, this tumor was non-metastasizing, but there were reports of multiple metastases in women treated initially by excision and who later succumbed to metastatic disease. About one-fourth of these tumors were pedunculated.[3,6]

There was no complete consensus regarding the tumor pathogenesis. This hormonally responsive tumor was believed to arise from specialized mesenchymal cells of the pelvic-perineal region or from the multipotent perivascular progenitor cells, which often displayed variable myofibroblastic and fibroblastic features.[7]

This hypothesis was supported by the fact that the tumor cells express desmin and in some cases, a smooth muscle actin. Recent cytogenetic and molecular studies had identified a variety of genetic alterations, involving the chromosome 12, in the region 12q.[13-15] A gene in this region, called high-mobility group protein isoform I-C (HMGIC), which encoded proteins involved in the transcriptional regulation, appeared to have a role in the pathogenesis of this tumor. Detection of inappropriate HMGI-C expression using the immunoperoxidase technique with anti HMGI-C antibody may potentially be a useful marker for microscopic residual disease.[8]

Microscopically, these myxomatous tumors were composed of spindle-shaped, stellate and plump oval cells in a myxomatous stroma. However, these myxomatous tumors showed different quantities and patterns of vessel elements.[9-11] The superficial angiomyxomas, both in the myxoid and collagen-fiber-rich areas, showed a scattered distribution of small to medium sized thin-walled blood vessels, but large caliber vessels, seen in aggressive angiomyxomas were absent. Moreover, a plexiform capillary pattern, detected in some malignant soft tissue tumors containing myxomatous lesions, such as low-grade fibromyxoid sarcoma, myxofibrosarcoma and myxoid malignant fibrous histiocytoma, was not seen.[12,13]

Clinically, AA may be misdiagnosed as Bartholin cyst, lipoma, labial cyst, Gartner duct cyst, levator hernia or sarcoma. Fibro-epithelial stromal polyp, superficial angiomyxoma, angiomyofibroblastoma, cellular angiofibroma and smooth muscle tumors also needed to be considered in the differential diagnoses of a polypoidal mass in the perineum. AA is an infiltrative tumor, whereas angiomyofibroblastoma is well circumscribed (this characteristic can also be identified on magnetic resonance imaging MRI). Also, AA had thick-walled vessels, which were less numerous than the thin-walled vessels in angiomyofibroblastoma.

On computed tomography (CT) scan, these tumors had a well-defined margin with attenuation less than that of the muscle. On MRI, these tumors showed high signal intensity on T2-weighted images. The attenuation on CT and high signal intensity on MRI were likely to be related to the loose myxoid matrix and high water content of angiomyxoma.[14]

Where fertility was to be preserved or surgery was likely to be extensive and mutilating, incomplete resection was acceptable as local recurrences can be treated with further resection. Recurrences may occur from months to several years after excision (2 months to 15 years).[14,15]

AA, despite the name, was not that aggressive, with only a 30% chance of recurrence, which was eminently treatable by excision with a 1 cm margin. Most of the patients had only one recurrence. Radiation therapy and chemotherapy were considered less-suitable options due to low mitotic activity. Hormonal manipulation with tamoxifen, raloxifene and gonadotropin-releasing hormone analogues had been shown to reduce the tumor size and may help to make complete excision feasible in large tumors and in the treatment of recurrence.[16]

Angiographic embolization may also helped in subsequent resection by shrinking the tumor as well as making it easier to identify it from surrounding normal tissues.[17]

As late recurrences were known, all patients needed to be counselled about the need for long-term follow-up. Magnetic resonance imaging was the preferred method for detecting recurrences.

CONCLUSION

Radiological studies and pathological evaluation aids in the diagnosis and planning of appropriate treatment of AA. Close and long term follow up of these patients should be emphasized due to high rate of local recurrence.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Elkattah R, Sarkodie O, Otteno H, Fletcher A. Aggressive angiomyxoma of the vulva: a précis for primary care providers. Case Rep Obstet Gynecol. 2013;2013:1-4.
2. Steeper TA, Rosai J. Aggressive angiomyxoma of the female pelvis and perineum. Report of nine cases of a distinctive type of gynecologic soft-tissue neoplasm. Am J Surg Pathol. 1983 [cited 2022;7(5):463-75.
3. Siassi RM, Papadopoulos T, Matzel KE. Metastasizing aggressive angiomyxoma. N Engl J Med. 1999;341(23):1772.
4. Kura MM, Jindal SR, Khemani UN. Aggressive angiomyxoma of the vulva: an uncommon entity. Indian Dermatol Online J. 2012;3(2):128.
5. Han-Geurts IJM, Geel AN, Doorn L, Bakker M, Eggermont AMM, Verhoef C. Aggressive angiomyxoma: multimodality treatments can avoid mutilating surgery. Eur J Surg Oncol. 2006;32(10):1217-21.
6. Blandamura S, Cruz J, Vergara L, Puerto I, Ninfo V. Aggressive angiomyxoma: a second case of metastasis with patient's death. Hum Pathol. 2003;34(10):1072-4.
7. Alameda F, Munné A, Baró T, Iglesias M, Condom E, Lloreta-Trull J, et al. Vulvar angiomyxoma, aggressive angiomyxoma, and angiomyofibroblastoma: an immunohistochemical and ultrastructural study. Ultrastruct Pathol. 2006;30(3):193-205.
8. Nucci MR, Fletcher CDM. Vulvovaginal soft tissue tumours: update and review. Histopathology. 2000;36(2):97-108.

9. Allen PW, Dymock RB, MacCormac LB. Superficial angiomyxomas with and without epithelial components. Report of 30 tumors in 28 patients. Am J Surg Pathol. 1988;12(7):519-30.
10. Bégin LR, Clement PB, Kirk ME, Jothy S, Elliott McCaughey WT, Ferenczy A. Aggressive angiomyxoma of pelvic soft parts: a clinicopathologic study of nine cases. Hum Pathol. 1985;16(6):621-8.
11. Fletcher CDM, Tsang WYW, Fisher C, Lee KC, Chan JKC. Angiomyofibroblastoma of the vulva. A benign neoplasm distinct from aggressive angiomyxoma. Am J Surg Pathol. 1992;16(4):373-82.
12. Evans HL. Low-grade fibromyxoid sarcoma. A report of two metastasizing neoplasms having a deceptively benign appearance. Am J Clin Pathol. 1987;88(5):615-9.
13. Goldblum J, Weiss S, Folpe AL. Enzinger and Weiss's Soft Tissue Tumors. 7th ed. Elsevier; 2019.
14. Outwater EK, Marchetto BE, Wagner BJ, Siegelman ES. Aggressive angiomyxoma: findings on CT and MR imaging. AJR Am J Roentgenol. 1999;172(2):435-8.
15. Behranwala KA, Thomas JM. Aggressive angiomyxoma: a distinct clinical entity. Eur J Surg Oncol. 2003;29(7):559-63.
16. McCluggage WG, Connolly L, McBride HA. HMGA2 is a sensitive but not specific immunohistochemical marker of vulvovaginal aggressive angiomyxoma. Am J Surg Pathol. 2010;34(7):1037-42.
17. Magtibay PM, Salmon Z, Keeney GL, Podratz KC. Aggressive angiomyxoma of the female pelvis and perineum: a case series. Int J Gynecol Cancer. 2006;16(1).

Cite this article as: Revanna R, Padmasri R, Divya TK. Aggressive angiomyxoma of uterine corpus. Int J Reprod Contracept Obstet Gynecol 2022;11:2023-6.