

## Introduction

Aggressive angiomyxoma (AAM) is a rare benign tumor, primarily affecting women of reproductive age, often in the perineal and pelvic regions. Although slow-growing, it has a high potential for local invasion. We present a case report of a woman with a second recurrence of a giant AAM originating from the labia majora.

## Discussion

AAM is a benign but locally aggressive tumor. While Aggressive angiomyxoma (AAM), first described in 1983, primarily affects women of reproductive age and commonly involves the perineum, vulva, vagina, pelvic cavity, and hips. It has a high recurrence rate (35-72%) within 2 years of resection. AAM tumor cells express estrogen and progesterone receptors, promoting growth during pregnancy. Its pathogenesis involves chromosomal translocation t(8;12) and changes in the 12q13–15 region.

Imaging studies like ultrasound and MRI are crucial for diagnosis and monitoring recurrence, with MRI providing superior detail. AAM has negative tumor markers (CA125, CEA, CA19-9), elevated Hsp90 levels linked to poor prognosis, and shows expression of vimentin, smooth muscle actin, desmin, CD34, F8, ER, and PR. Treatment requires complete surgical resection with negative margins, and adjunct therapies such as gonadotropin-releasing hormone agonists or ER/PR-targeted therapies may help. Long-term follow-up for up to 15 years is recommended due to high recurrence and rare metastasis.

## Case Description

A 34-year-old female presented with a progressively enlarging mass on the right labia majora, causing perineal heaviness and abstinence due to discomfort. She had a history of surgically treated aggressive angiomyxoma (AAM) in 2013 and 2015. Examination revealed a soft, pedunculated mass (20×12 cm) (**Fig 1**) with no discharge or inguinal lymphadenopathy. Ultrasound showed a well-defined, heterogeneous hyperechoic lesion (12.2×12.1×5 cm), and MRI revealed a hyperintense, well-defined soft tissue mass (19×10×8.7 cm) without adjacent infiltration.

## Conclusion

AAM is a benign but locally aggressive tumor. While physical examination and imaging aid in diagnosis, HPE is confirmatory. En bloc excision with negative margins is recommended to prevent recurrence, and long-term annual follow-up with MRI is advised.

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## Procedure

The patient underwent wide local excision of a 25×17×12 cm pedunculated mass from the right labia majora, attached by a 5×5×5 cm peduncle. The stalk was identified, clamped, cauterized, and the mass removed en bloc for histopathology. The mass appeared yellowish-white, and HPE confirmed recurrent aggressive angiomyxoma (AAM) (**Fig. 2A and B**). with spindle, round, and stellate cells in a myxoid stroma and hyalinized blood vessels. The patient has been recurrence-free for 17 months post-surgery.

## References

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**Conflict of Interest** - None declared.

