



Research Article

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## Aggressive Angiomyxoma Clinical Images

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### Abstract

Aggressive angiomyxoma (AAM) is a rare, commonly infiltrative mesenchymal tumor presenting mainly in the inguinal and perineal region of reproductive-age females, between the second and fourth decades of life, with vague symptoms. Surgical resection is the treatment. We describe a case of a perineal aggressive angiomyxoma in a 44-year-old woman in which the diagnosis was made after histological examination. Close long-term follow-up by MRI is mandatory. Most recurrences are local, within the first 3 post-operative years. There are no reports of AAM as cause of death, although multiple-recurrence cases can impact quality of life.

### Keywords

Angiomyxoma; Perineal neoplasm; Inguinal neoplasm

### Case Presentation

A forty-four-year old woman was seen in General Surgery clinic with complaints of discomfort after prolonged sitting, and aching in her left buttock evolving over several months. Her medical history included smoking, hypercholesterolemia and conization in 1997. Colonoscopy, in 2011, was unremarkable. She was pre-menopausal. On physical examination, there was no palpable or visible buttock mass and the digital rectal examination was unremarkable.

Pelvic MRI (Figures 1 and 2) revealed an ovoid lesion, 6.5 cm × 5.5

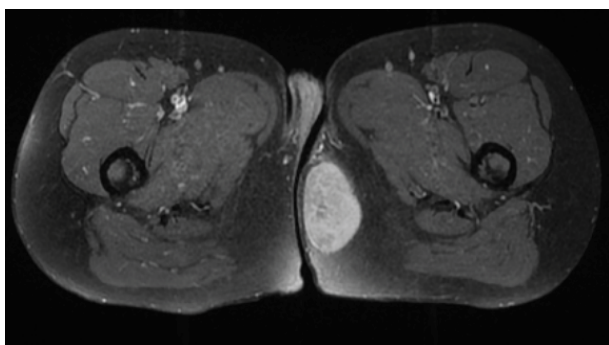


Figure 1: MRI axial T1 gadolinium fat saturation. Ovoid lesion in the adipose tissue of the left inferior ischioanal fossa, enhanced intensely after injection.

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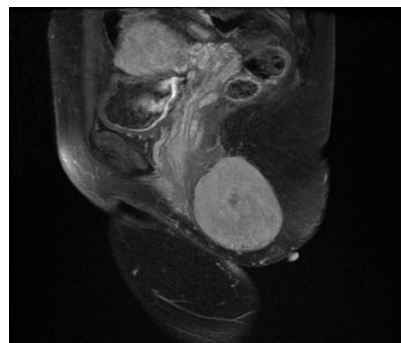


Figure 2 : MRI Sagittal T1 gadolinium: lesion with his vascular pedicle.

cm × 3.7 cm, in the adipose tissue of the left inferior ischioanal fossa. It enhanced intensely after injection, appeared iso-dense to muscle on T1 and discretely hyper-intense on T2-weighted images. There was no invasion of surrounding fat or thickening of the rectal wall. The lesion was in direct contact with levator ani muscle causing subtle distortion of the anal canal, but without evidence of musculotendinous invasion.

Our surgical intervention consisted of complete resection of the tumor by a perineal approach. The tumor was in direct contact with the rectum, vagina and levator ani muscle on the left. Postoperative histopathology revealed amitotic spindle cells, confirming the diagnosis of AAM. In anatomopathological examination, the tumor had elastic consistency and was pinkish beige, limited by a pseudocapsule. There were no necrosis and no infiltration of the muscle partially resected. Ki 67 was estimated to 5 % and estrogen receptor and progesterone receptor were expressed. The resection was RO although tangential, wide margins were not sought due to associated morbidity. Postoperative recovery was smooth with no apparent complications at one year follow-up. Our patient will be closely followed with repeat MRI every six months for the first two years.

### Discussion

Around 250 cases have been reported in the literature since 1983, forty in men within the same anatomic region. Female-to-male ratio is approximately 6:1 [1].

Because of its rarity, location and nonspecific clinical presentation, misdiagnosis is common [2-5]. Without imaging, the extent of these tumors is often not appreciated until surgery. Our patient had pain after prolonged sitting, we did not feel the mass by digital examination while MRI revealed a 6,5 cm tumor. Although histopathology is the gold-standard, MRI is useful and reveals T1 hypointensity, T2 hyperintensity, avid heterogenous enhancement after contrast administration and a distinct, low-intensity, swirling pattern [6]. Preoperative diagnosis by MRI with biopsy confirmation [7] is crucial for surgical planning [1-3,6,8].

Histology reveals acellular, myxoid and hypervascular characteristics with frequent estrogen and progesterone receptor over-expression. Tumors are frequently greater than 10 cm, indolent, with locally aggressive behavior and high rates of recurrence (27% -

70% in small series) despite R0 resection.

Multiple studies (although small) suggest that recurrence rates are the same whether R0 or R1 resection is obtained [2]. As such, wide local excision for small masses is advocated, but for larger masses en bloc resection is discouraged if significant post-operative morbidity will result [1,4,9]. If estrogen or progesterone receptor positive, neoadjuvant hormonal therapy (with a GnRH agonist) may be useful, although evidence is mainly case reports and small series [10]. Close long-term follow-up by MRI is mandatory. Most recurrences are local, within the first 3 post-operative years. There are no reports of AAM as cause of death, although multiple-recurrence cases can impact quality of life. Metastases are exceedingly rare.

### Conclusion

Aggressive angiomyxoma should be considered in the differential diagnosis of premenopausal women who present with a pelvic mass. MRI can assist in non-invasive diagnosis and personalized pre-operative planning emphasizing preservation of function. Confirmatory diagnosis is with histopathology. Long-term MRI follow-up is mandatory. Prognosis is good.

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