Aggressive Angiomyxoma of the Vulva or Perineum: Report of Three Patients

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Abstract

Background: Aggressive angiomyxoma is a rare, locally aggressive soft tissue tumour that chiefly involves the vulvoperineal region of young female patients. Treatment is wide surgical excision. Frequent relapses are common.

Cases: We present three patients who each had an aggressive angiomyxoma treated initially for another presumed diagnosis. Two patients required immediate repeat surgical procedures for incomplete tumour excision. The third patient developed a recurrence two years after the initial surgery. In one patient, hormonal therapy was used postoperatively because the evaluation of the tumour margins was uncertain. Harpoon markers were used to aid in tumour localization in another patient prior to resection.

Conclusion: Clinicians should consider the diagnosis of aggressive angiomyxoma when a patient presents with an atypical vulvoperineal mass, as an incorrect diagnosis may lead to repeated surgical procedures.

Résumé

Contexte : L'angiomyxome agressif est une tumeur rare, localement agressive, du tissu mou qui affecte principalement la région vulvopérinéale de jeunes patientes. Le traitement consiste en une excision chirurgicale large. De fréquentes récidives sont courantes.

Cas : Nous vous présentons le cas de trois patientes qui présentaient toutes un angiomyxome agressif ayant été initialement pris en charge en fonction d'un diagnostic présumé erroné. Deux patientes ont nécessité immédiatement une deuxième intervention chirurgicale en raison d'une excision incomplète de la tumeur. La troisième patiente a présenté une récidive deux ans après la chirurgie initiale. Chez l'une des patientes, nous avons eu recours à l'hormonothérapie à la suite de l'opération en raison du caractère incertain de l'évaluation des bords de la tumeur. Des marqueurs « *harpoon* » ont été utilisés, avant la résection, pour faciliter la localisation de la tumeur chez une autre patiente.

Conclusion : Les cliniciens devraient envisager le diagnostic d'angiomyxome agressif lorsqu'une patiente présente une masse vulvopérinéale atypique, puisqu'un diagnostic erroné peut mener à des interventions chirurgicales répétées.

Key Words: Aggressive angiomyxoma, recurrence, harpoon markers

Competing Interests: None declared.

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INTRODUCTION

Aggressive angiomyxoma is an uncommon soft tissue tumour that was first described in 1983 by Steeper and Rosai.[1] It is a slow-growing, locally destructive tumour.[2] It is generally sex-steroid receptor positive.

The tumour usually presents in women in the reproductive age group as a perineal or vulvar mass. Treatment is wide surgical excision. Local recurrences are frequent.[3]

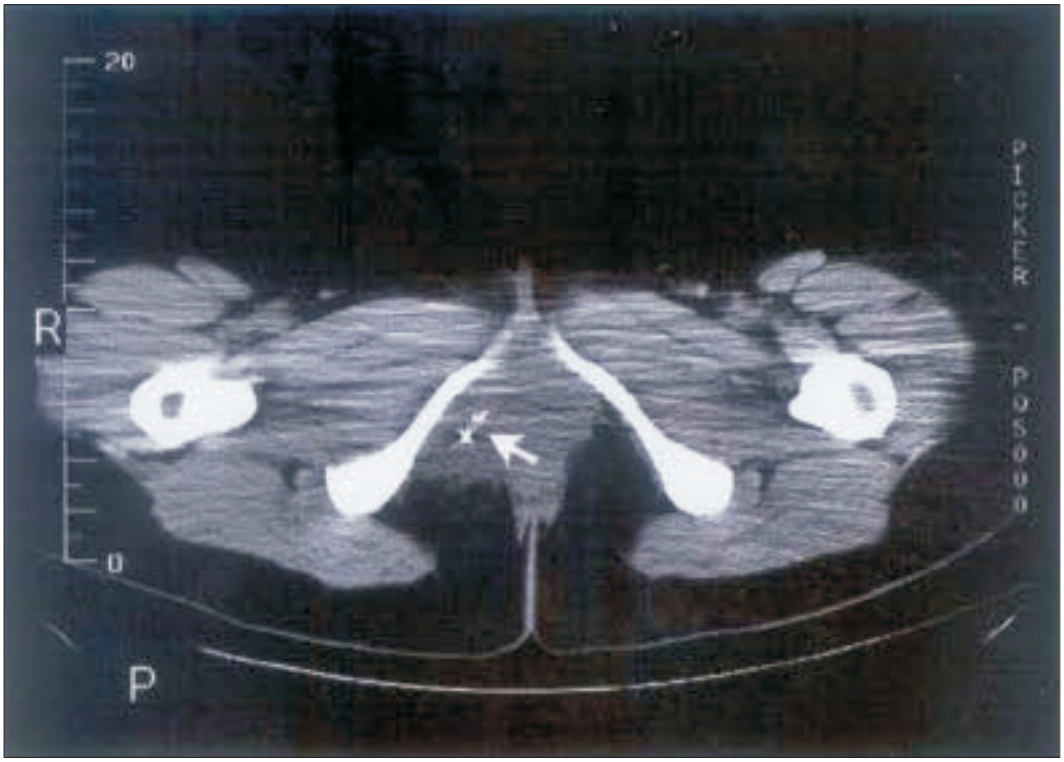
We report here three cases of aggressive angiomyxoma, none of which had this histologic diagnosis suspected preoperatively. As a result, two patients required further surgical intervention because of incomplete initial surgical excision.

THE CASES

The first patient, a 51-year-old woman, had a history of resection of a left-sided vulvar mass at age 45 for a presumed Gartner's duct cyst. The pathological diagnosis at that time was fibroadenoma. Two years later she had recurrence of a left-sided vulvar mass that was resected surgically. The pathology report described aggressive angiomyxoma. The resection margins were free of tumour. A review of the histology from the first procedure indicated that the primary tumour had been aggressive angiomyxoma. At a follow-up visit six years after the initial diagnosis, she remained free of recurrence.

The second patient, a 41-year-old woman, complained of non-specific pelvic pain that had been present for several years. The patient discovered a right vulvar mass that was slowly growing. She underwent surgery for a presumed diagnosis of perineal hernia. The mass was infiltrating the perineal muscles, and the surgeon felt the resection was incomplete. The histology of the resected mass showed aggressive angiomyxoma. Postoperative magnetic

Figure 1. The 4cm right paravaginal mass with ill-defined borders identified by two harpoon markers (arrow).



resonance imaging (MRI) showed a 5-cm residual mass in the right ischiorectal fossa.

The mass was not palpable, so two harpoons were inserted preoperatively under computerized tomography (CT) guidance to help localize the tumour (Figure 1). The resection was carried out in the region indicated by the harpoons. The histology of the resected mass confirmed persistent aggressive angiomyxoma. Resection margins were free of tumour. Postoperative X-ray films confirmed removal of the inserted markers. The patient remained free of disease 30 months after surgery.

The third patient, a 30-year-old woman, presented with a right-sided vulvar mass. She underwent surgery, performed by a general surgeon, for a presumed diagnosis of perineal hernia. Intraoperatively, she was found to have a multilobular mass originating from the right labium majus and extending retroperitoneally. The excision required an abdominoperineal approach, as the surgeon was not able to excise the mass through a perineal incision. The excised mass weighed 1.6 kg. Histological examination confirmed the diagnosis of an aggressive angiomyxoma (Figure 2). Postoperative MRI revealed a persistent 4.6cm mass in the right ischiorectal fossa. The patient therefore had further surgery performed by a gynaecologic oncologist, with wide local excision. The surgical margins were difficult to assess pathologically because the tissue specimen was fragmented.

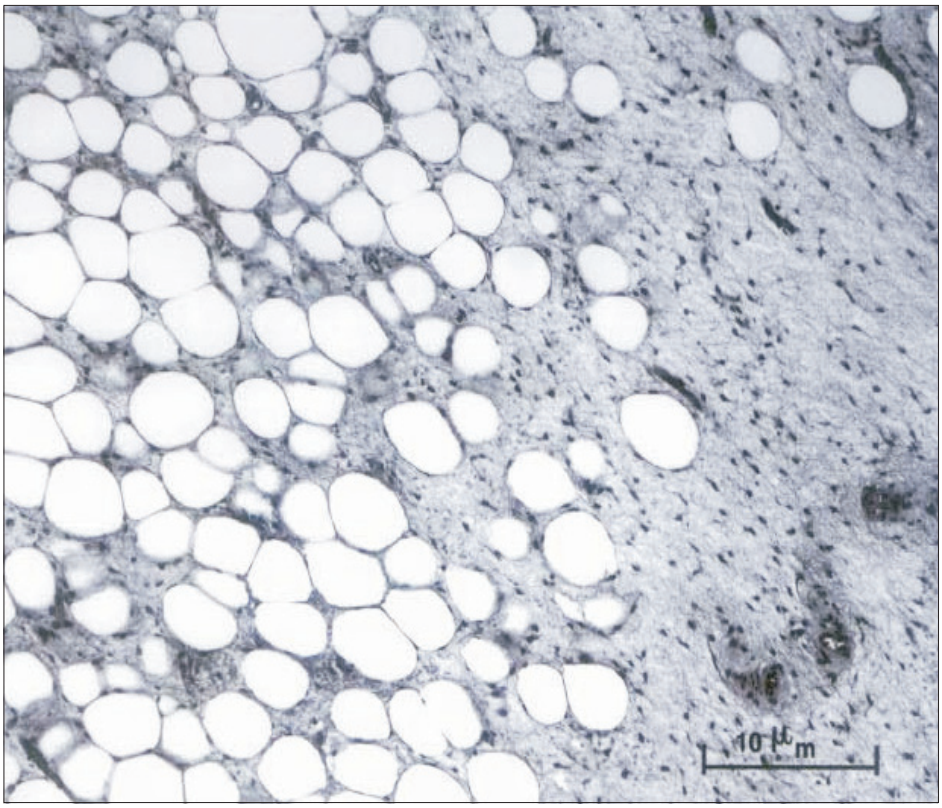
The tumour was estrogen and progesterone receptor-positive. The patient was given three injections of a gonadotropin-releasing hormone (GnRH) agonist at monthly intervals as postoperative adjuvant therapy. She remained free of disease nine months after the second surgical procedure.

Each patient was seen in follow-up at three to four month intervals for the first two years, and every six months thereafter. MRI was performed every year for the first two years after surgery, or more often if thought to be advisable.

DISCUSSION

In 1983, Steeper and Rosai first described aggressive angiomyxoma as a distinctive variant of myxoid neoplasms that had a predilection for the pelvis and perineum in young female patients.[1] It occurs less frequently in males: the female-to-male incidence ratio is 6:1. The peak incidence is at 35 to 40 years of age (range 6–77).[3] Begin et al. subsequently described nine cases of aggressive angiomyxoma, all characterized by slow insidious growth of a focally

Figure 2. Microscopic appearance of aggressive angiomyxoma, showing distinctive myxoid appearance and spindle-shaped fibroblasts.



infiltrative tumour with no capsule. All tumours recurred locally and none metastasized. Recurrences were attributed to incomplete tumour excision.[2]

The recurrence rate of these tumours is high, in the range of 36% to 72%.[1–3] Two recent reviews reported a very low recurrence rate (9%), although the authors of these reviews commented that because recurrence might occur several years after surgical resection, this low figure might be due to a short period of follow-up.[4,5] Most recurrences occur locally, but two recent case reports documented distant metastases resulting in the death of one of the patients.[6,7] Htwe reported a case where growth was apparently related to pregnancy. Because the neoplasm was strongly progesterone-receptor positive, the tumour was thought to be hormone-related.[8]

Ninety percent of patients with aggressive angiomyxoma are in the reproductive age group. In a review by Fetsch et al., the peak incidence in females was in the fourth decade of life. In this review, evidence of estrogen receptors in the tumour was found in 13 out of 14 cases and progesterone receptors in 9 of 10 cases.[3]

A report of successful adjuvant therapy with a GnRH agonist in a patient following complete surgical excision of an aggressive angiomyxoma[9] was followed by the first report of primary medical management of a patient with recurrent aggressive angiomyxoma of the vulva using a GnRH agonist.[10] The latter patient had complete resolution following treatment. She did not require further medical or surgical intervention.

Misdiagnosis has been reported in 82% of cases.[11] In our series, two patients had initial procedures performed by general surgeons for a presumed diagnosis of perineal hernia. In both cases the surgeons were faced with a deeply infiltrating tumour, and complete resection was not achieved. Both patients required a repeat surgical procedure performed by a gynaecologic oncologist in order to excise the tumour mass completely.

Adjuvant hormonal therapy was given to the third patient in our series because it was difficult to determine whether the tumour was completely excised. The tumour contained both estrogen and progesterone receptors. The patient was treated with three monthly injections of a GnRH agonist,

following previous reports of favourable outcomes with such treatment in the adjuvant and primary treatment settings.[7–8] We elected to treat this patient for only three months to avoid the adverse effects of prolonged GnRH agonist use. Nine months after completing treatment, the patient remained free of recurrence.

Tumour localization may be difficult after incomplete surgical excision or in cases of early recurrence that are evident only with use of imaging. We found the use of harpoon markers inserted under CT guidance to be useful for tumour localization and as a guide for surgical excision.

CONCLUSION

Clinicians should consider the diagnosis of aggressive angiomyxoma when a patient presents with an atypical vulvoperineal mass, as an incorrect diagnosis may lead to repeated surgical procedures. If a patient presents with a vulvoperineal mass with an unusual location or texture, a complete physical examination (including rectal examination) may show the infiltrative nature that is indicative of these tumours. Studies using CT or MRI may be of use in demonstrating the locally invasive nature that typifies these tumours. Preoperative fine needle biopsy may also be useful. The differential diagnosis of such masses includes vulvar abscess, Gartner's duct or Bartholin's gland cyst, perineal hernia, vaginal polyp, lipoma, hematoma, and carcinoma of the vulva or Bartholin's gland.

Because these tumours tend to recur locally and early recurrences are difficult to diagnose, we use yearly MRI studies postoperatively to rule out early recurrences.

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