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Recurrence of an Aggressive Angiomyxoma of the Pelvis: Case Report

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Abstract **Case Report**

Aggressive angiomyxoma is a rare, slow-growing mesenchymal tumor of vulvo-perineal and pelvic topography in women of reproductive age. We report the case of a 33-year-old female patient in the genitally active period who underwent surgery for pelvic pathology and presented with chronic pelvic pain with a sensation of heaviness, evolving in a context of apyrexia and preservation of general condition. The anatomopathological diagnosis was aggressive pelvic angiomyxoma associated with a recurrence of her disease. The diagnosis of aggressive angiomyxoma should be made in a genitally active woman presenting with a slow-growing mass of the pelvis and perineum. Preoperative diagnosis using MRI will alter the patient's surgical management and prognosis.

Keywords: Aggressive angiomyxoma, pelvic topography, chronic pelvic pain.

INTRODUCTION

Aggressive angiomyxoma is a rare, slow-growing mesenchymal tumour of vulvo-perineal and pelvic topography in women of childbearing age [1, 2]. First described in 1983 by Steeper and Rosai [3], less than 350 cases have been reported in the literature. Treatment is surgical [4].

Clinical diagnosis is often unrecognized [5, 6], yet preoperative knowledge of the diagnosis modifies surgical modalities, and the patient's prognosis; recurrence is frequent in cases of incomplete excision [6]. Recognition of this typical but rare lesion on MRI is therefore essential.

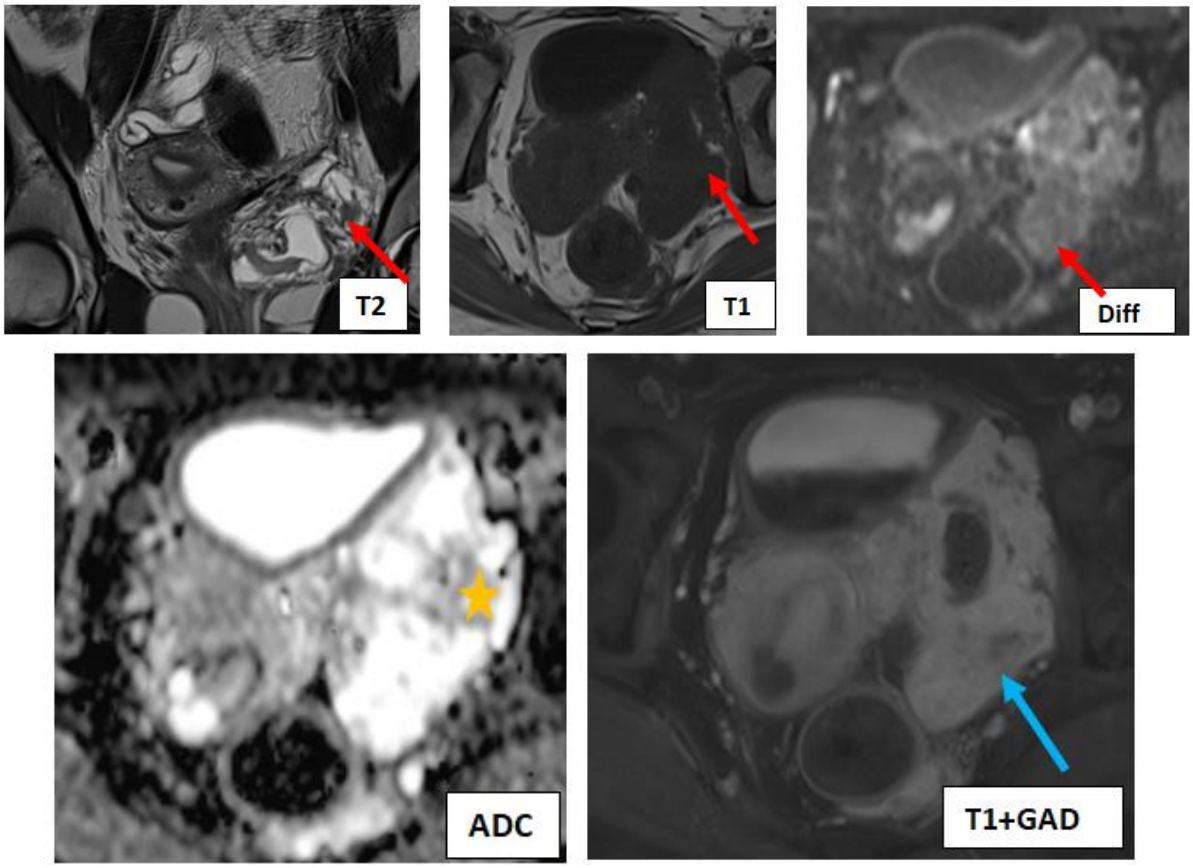
We report the case of a 33-year-old woman presenting with an aggressive pelvic angiomyxoma of para-vaginal para-rectal localization, whose initial diagnosis, although specific on imaging, was not recognized.

OBSERVATION

A 33-year-old female patient in good general condition, initially operated on for a pelvic pathology, now presented with chronic pelvic pain with a sensation of heaviness. A pelvic MRI was performed, revealing: a left para-vaginal and rectal lesion with poly-lobed contours, fairly well limited, in T1 hyposignal, T2 heterogeneous hypersignal, intensely and heterogeneously enhanced after injection of PDC, containing areas in diffusion hypersignal with ADC restriction.

This process comes into contact with the lateral wall of the cervix and vagina medially, as well as the middle and lower rectum, respecting the separating fatty border, and also infiltrates the homolateral parametrium. Posteriorly, it extends retro-rectally, respecting the separating fatty border. Further down, it fills the pre-rectal space and infiltrates the perineal floor; the laboratory work-up is unremarkable.

The anatomopathological diagnosis was that of an aggressive benign angiomyxoma. The post-operative course was straightforward.



Pelvic MRI with weighted sequences: T2, T1, diffusion, ADC and T1 with gadolinium injection in axial and coronal sections: Left pararectal and vaginal lesional process (red arrow) with poly-lobed contours, fairly well limited, in T1 hyposignal, heterogeneous T2 hypersignal, intensely and heterogeneously enhanced (blue arrow), containing areas in diffusion hypersignal with ADC restriction (star)

DISCUSSION

Aggressive angiomyxoma is a rare mesenchymal tumour, occurring in 90% of cases in women of childbearing age [1, 5, 7], usually between the third and fourth decade [8]. Very rare cases have been reported in men, with identical inguino-scrotal and perineal topographies [3, 9].

Clinically, due to its slow, insidious growth in an extra-compartmental situation (with no anatomical limit to its development) [1], and its "flabby" consistency, patients are often asymptomatic [6], the lesion being discovered once it has become voluminous. When the tumor is symptomatic, clinical symptoms frequently include pelvic heaviness, vulvovaginal pain, dysmenorrhea, dyspareunia, digestive and urinary disorders [1, 8].

True extension is largely underestimated due to the fact that it extends above the pelvic diaphragm [1].

On ultrasonography, aggressive angiomyxomas appear hypoechoic or even pseudocystic [7, 10], with only the vascularization, demonstrated in Doppler mode, confirming their tissue nature.

On CT, the tumor appears spontaneously hypodense in relation to the muscle, or even pseudocystic [11].

On MRI, the tumour appears in T1 hyposignal and intense T2 hypersignal. Spontaneous hypodensity on CT and intense T2 hypersignal reflect the water-rich, myxoid nature of the lesion. After injection of gadolinium, it produces a swirled or striated appearance, appearing as hyposignal lines within the enhanced tumor.

This sign, described as typical in the literature [1, 10, 11], can also be found on T2 and CT scans, particularly after injection [7]. Although incompletely elucidated, this aspect could reflect stretching of the fibrovascular stroma on either side of the pelvic diaphragm [11].

On both CT and MRI, the tumour typically appears as a well-limited mass, pushing back the neighbouring organs without infiltrating them (urethra, vagina, anal sphincter and rectum). These two techniques can be used to determine the extent of the tumour, on either side of the pelvic diaphragm, which determines surgical planning.

However, MRI remains the best tool, given its better contrast resolution in the pelvis. Post-operatively, it can be used to search for macroscopic remnants, and to monitor for distant recurrences, which will have the same characteristics as the initial tumour [1, 6, 7].

The combination of the patient's condition (female genital activity), the topography of the lesion and the MRI appearance (well-defined mass with a myxoid signal) make the diagnosis virtually certain.

On clinical examination, the diagnosis is often overlooked because it is confused with more common diagnoses such as a Bartholin's gland cyst, abscess or hernia, all of which are easily confirmed on MRI [6, 8]. In anatomopathology, the differential diagnoses are benign lesions: superficial angiomyxoma, cellular angiofibroma, myxoid leiomyoma or even deep abdominal fibromatosis (desmoid tumour), or malignant lesions such as myxoid liposarcoma.

A recent study demonstrated the value of HMGA2 immunohistochemical expression testing as a potential diagnostic aid [15].

In their study, 90% of aggressive angiomyxomas were nuclear positive, whereas benign genital stromal tumours were nuclear negative. However, 27% of fibroepithelial stromal polyps were positive, as were 33% of leiomyomas.

Treatment is surgical, involving excision in a single block. Radiotherapy is not very effective, due to its low mitotic activity [18].

Hormone therapy with a Gn-RH analogue may have a place as primary treatment for small, pauci-symptomatic tumours, or as adjuvant therapy for tumour remnants and recurrences [1].

When excision is incomplete, recurrence occurs in 9 to 73% of cases, depending on the series [1], which is why the tumor is described as "aggressive" [1, 5], despite being benign.

This high rate of local recurrence is due to a failure to recognize the diagnosis preoperatively (this tumour has a tendency to haemorrhage in the event of invasion, which is favoured by the absence of a capsule) and to an underestimation of deep extension (above the pelvic diaphragm, sometimes requiring a double approach [1, 7]), local recurrence being correlated with the surgeon's experience.

Despite the morbidity associated with recurrence and the sometimes iterative surgical procedures, the prognosis for aggressive angiomyxoma remains good overall [1]. Only two cases of peritoneal implants have been reported in the literature [19, 20]. Clinical and MRI follow-up is recommended [8].

CONCLUSION

Aggressive benign angiomyxoma is a rare pathology, which the radiologist must consider when faced with a recurrent soft, mobile and often painless tumour in the para-vaginal or para-rectal space in a young woman.

The diagnosis of certainty is anatomopathological, based on ultrasound-guided perineal biopsies. This tumour must be managed in a setting specialising in soft tissue tumours, to avoid local recurrence.

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