RADIOLOGIC PATHOLOGIC CORRELATION / Genito-urinary imaging

Aggressive angiomyxoma

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Gonadotropin releasing hormone (GnRH) agonists are used in the treatment of hormone-dependent gynaecological tumours.

The authors report the case of a patient treated for MRI-diagnosed and monitored, multi-recurrent aggressive angiomyxoma, a rare disease that regressed under Enantone\textsuperscript{a} (Leuprelin).

Case report

In 2002, a young 36-year-old woman underwent surgery for a 3 cm by 8 cm tumour on the recto-vaginal septum, responsible for invalidating pelvic pain (Fig. 1a and b). The excision was complete and the diagnosis was that of atypical leiomyoma.

Five years later, in January 2007, the patient presented a recurrence of the symptoms. The MRI detected a local tumour of 11 cm located at the site of the initial excision (Fig. 1c and d). The patient again underwent surgery (Fig. 2a). The anatomopathological diagnosis was changed to that of aggressive angiomyxoma (Fig. 2b and c). The histology revealed the presence of tumour cells expressing œstrogen and progesterone receptors (Fig. 2d).

In June 2008, 18 months after the second intervention, another local recurrence was noted (Fig. 3a and b). In view of the hormone-dependent nature detected in the previous histology, a decision was made to avoid another intervention and start treatment with Enantone\textsuperscript{a} (a GnRH agonist). The lesion regressed 6 months after beginning treatment. Only a fibrous strip was visible in the imaging (Fig. 3c and d).

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Eighteen months later, the patient suspended the treatment due to the poorly tolerated adverse effects. The lesion returned 6 months after the suspension of the treatment (Fig. 4a and b). It regressed as soon as the treatment was restored (Fig. 4c and d). The patient is currently completely stabilised by this treatment and a distant location has not been observed.

**Discussion**

Aggressive angiomyxoma is difficult to diagnose because it is rare (described in 1983, about 250 cases in the literature), although knowledge of its radiological semiology [1], the predisposition and its natural history should call it to mind.

This myxoid mesenchymateous lesion of the perineal pelvic region is locally aggressive although benign (only two cases of secondary locations have been reported). Ninety percent of the cases involve young women in their 4th decade. Several cases have been described in men, with scrotal and inguinal locations, and a peak at 60 to 70 years.

The clinic picture is aspecific, characterised by the often asymptomatic, slow growth of a soft and mobile mass in the paravaginal or pararectal space. Initially, the lesion is erroneously taken for a leiomyoma or a parasitic myoma [2]. It differs from diffuse peritoneal leiomyomatosis due to its extra-peritoneal location, or a rectal GIST, a very rare location and the existence of a healthy wall and a border of fat separating the lesion from the rectal muscularis. Retorectal tumours are not found in this place and specifically present a liquid or fatty sheath although heterogeneous [3].

In MRI, the lesion appears well limited in hypersignal T2, hyposignal T1, taking the contrast heterogenically. The typically paravaginal or pararectal location and its very specific local extension indicate this diagnosis and allow it to be distinguished from retro-rectal tumours.

Macroscopically, the lesion appears as a soft, gelatinous, non-encapsulated mass without distinct limits, with a polylobed outline, infiltrating the adjacent soft tissue.

In histology, this lesion consists of connective cells dispersed on a myxoid and collagen background with rich vascularisation. There is no cytonuclear atypical or mitosis. The tumour cells express the hormone receptors to oestrogens and progesterone, thereby confirming the hormone-dependent nature of this lesion [4].

Therefore, the use of GnRH agonists, such as leuprorelin (Enantone®), is justified to inhibit the pituitary secretion of gonadotropins (FSH/LH). Leuprorelin fixes on the same liaison site as endogenous GnRH on its receptor, but with a higher affinity and an inhibiting effect.
Figure 2. Macroscopy and histology. a: macroscopic sample of the recurrence in 2007; b: HES, × 40, HES staining: proliferation or small fusiform or star-shaped cells, without cytonuclear atypia (arrow) dispersed on a myxoid background (solid arrow) enclosing several capillary structures with a thin wall; c: HES, × 40, immunohistochemistry: rare smooth residual muscle cells stained by anti-smooth muscle antibodies (arrow). Absence of staining of the tumour cells; d: × 40, immunohistochemistry: coloration of most of the nuclei by anti-oestrogen antibodies (arrow).

In the case of our patient, the resulting state of hypogonadism ends by a spectacular reduction in the size of the tumour, although complicated by the classic adverse effects of menopause (hot flashes, sweating at night, mood disorders).

The specific diagnosis of this tumour and its hormone-dependent nature enable medical treatment and avoid mutilating surgery, that is, abdominal-perineal amputation in young women, without any effect on the number of recurrences [5,6]. The several cases in the literature also reveal a satisfactory response in carcinological terms under long-term treatment [7,8].

The originality of this case lies in the efficacy of Enantone® with regression in the size of the tumour as of 6 months of treatment. The premature suspension (after 18 months) of this treatment, due to the adverse effects, very quickly led to a tumour recurrence that regressed as soon as the treatment was resumed. This also demonstrates the direct response and on/off effect of hormone therapy on this locally aggressive lesion.

Due to the hormone-dependence of this lesion, the patient will therefore continue treatment until physiological menopause [9], associated with monitoring and the preventive treatment of osteoporosis.
Figure 3.  a, b: MRI of the second recurrence, June 2008; a: axial plane T1 after injection and saturation of the fat; b: frontal plane T2: appearance of two tissue nodules on the fibrous scar, 6 mm and 14 mm (arrows), presenting the same characteristics as the initial lesion. Establishment of a hormone treatment (Enantone®); c, d: MRI carried out 6 months after beginning the hormone treatment, December 2008; c: axial plane T1 after injection and saturation of the fat; d: frontal plane T2: regression of the tumour taking on the appearance of a fibrous strip (tip of the arrow).
Figure 4. a, b: MRI of the third recurrence, January 2011, 6 months after the suspension of Enantone®; a: axial plane T1 after injection and saturation of the fat; b: frontal plane T2: nodular lesion (arrows) with homogenous and intense contrast, in hypersignal T2, opposite the right anterolateral edge of the anal canal and lower rectum, measuring 25 × 34 × 52 mm, in contact with the posterior wall of the vagina. Resumption of the hormone therapy; c, d: MRI carried out 1 year after the third recurrence and resumption of the hormone therapy, January 2012; c: axial plane T1 after injection and saturation by the fat; d: frontal plane T2: regression of the fibrous, cicatricial tumour (tip of the arrow). The patient is still receiving hormone therapy.

Conclusion
Aggressive angiomyxoma is a rare disease that the radiologist should keep in mind when confronted with a soft and mobile, often painful, recurrent tumour of the paravaginal or pararectal space in a young woman. Treatment with a GnRH agonist avoids mutilating surgery that does not prevent recurrences.

Disclosure of interest
The authors declare that they have no conflicts of interest concerning this article.

References