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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® (Leuprorelin).

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 symptomatology. 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® (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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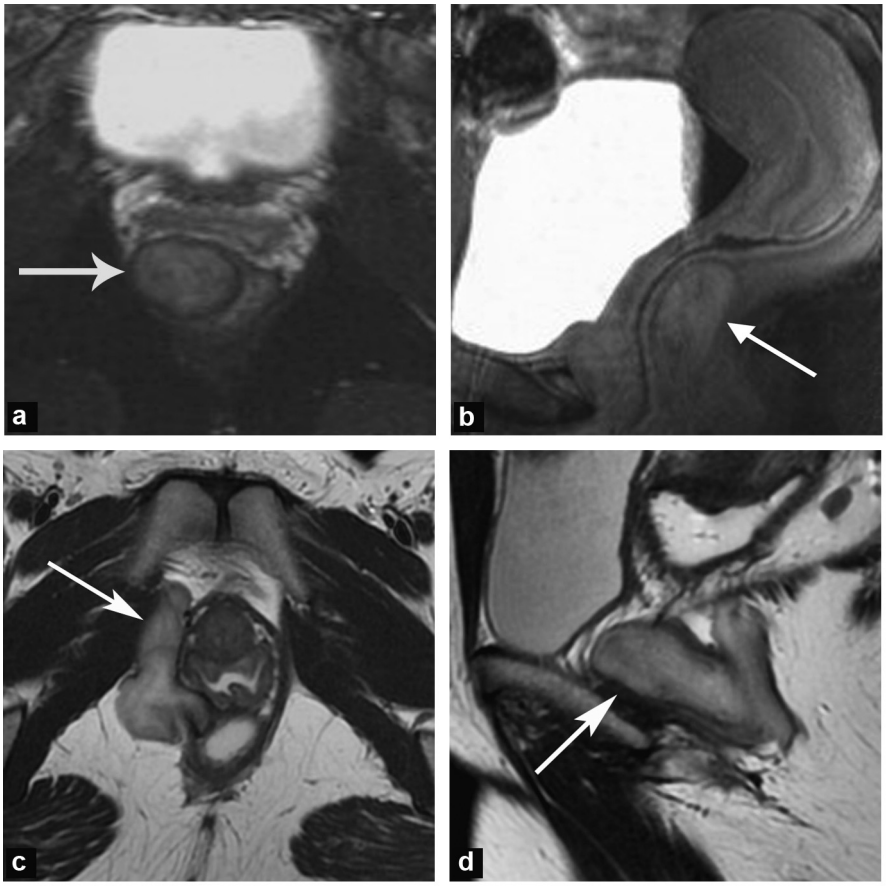

Figure 1. a, b: MRI of the initial lesion, 2002; a: axial plane; b: sagittal plane weighted sequence T2: tumoral lesion (arrow) of the recto-vaginal space, 48 × 22 × 20 mm, heterogeneous in moderate hypersignal T2; c, d: MRI of the first tumoral recurrence, 2007; c: axial plane; d: sagittal plane, weighted sequence T2: tissue lesion of the recto-vaginal space (arrow) extending along the right lateral wall of the vagina, in contact with the internal anal sphincter, the right puborectalis muscle is no longer visible.

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. Retrorectal 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 atypy or mitosis. The tumour cells express the hormone receptors to œstrogens 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.