Vulvar painless mass in a 41-year-old woman

Published on 07.08.2019

DOI: 10.35100/eurorad/case.16424
ISSN: 1563-4086
Section: Genital (female) imaging
Area of Interest: Genital / Reproductive system female
Imaging Technique: MR
Case Type: Clinical Cases
Authors: Elena Zabía Galíndez1, Maria Isabel Mata2, Elena Montoro2, Patricia Calvo2
Patient: 41 years, female

Clinical History:

We report the case of a 41-year-old woman with a left-sided vulvar mass which she had had for the past three years with very slow growth and without any pain, skin changes, discharge or bleeding.

Imaging Findings:

MRI examination revealed a large soft-tissue mass, mainly located in the left perineum and extending to the perirectal space. The mass displaced the adjacent soft tissue and split the internal and external right elevator ani sphincter muscle. It was hyperintense to muscle on T2-weighted images (Fig. 1), isointense to muscle on T1-weighted images (Fig. 2) and showed a whirling pattern and avid contrast enhancement (Fig. 3). Surgery confirmed a pedunculated partially encapsulated soft tissue mass extending through the pelvic diaphragm displacing adjacent structures without infiltration. (Fig. 4)

Discussion:

Aggressive angiomyxoma is a very rare soft tissue mesenchymal tumour described for the first time in 1983 [1], with only about 350 cases in the literature [2]. It usually occurs in the vulvovaginal region, pelvis and perineum of women in the reproductive age but has been described in the inguinoscrotal region of men in older ages [3], in children and in perimenopausal women [4]. The female-to-male ratio is about 6:1 [5]. It is a benign tumour called “aggressive” because of its 25% to 47% postsurgical local recurrence rate [3]. Metastases have been described in three cases, in the lungs or mediastinum [6-8]. It grows slowly as a painless solid mass with unclear borders around the perineal and pelvic structures and can occupy the whole pelvic cavity. [9] Despite extensive tumour both above and below the pelvic floor and even if the tumour infiltrates the pelvic organs or muscles, patients have few symptoms related to urinary, sexual or anal sphincter dysfunction. [4, 10] and usually present with a painless mass or with discomfort from pressure effects on adjacent pelvic organs. Due to its nonspecific clinical presentation and its rarity, aggressive angiomyxomas can be easily misdiagnosed as a Bartholin’s or Gartner’s duct cyst, a hernia, an abscess, a lipoma and other soft tissue tumours or even as a malignant pelvic floor tumour [11]. Macroscopically the surface of the tumour is smooth and partially or completely encapsulated. Microscopically the
tumour is composed of thick-walled vessels in a loose collagenous and myxoid stroma with spindle and stellate cells, generally with low mitotic activity, no cytologic atypia and no cell necrosis. [3]

Angiomyxoma has a characteristic MR appearance related to its loose myxoid matrix, high water content and high vascularity: It is usually hyperintense to muscle on T2-weighted images), iso or hypointense to muscle on T1-weighted images (Fig. 2) and avidly enhances after contrast administration (Fig. 3), with typical swirling and layering patterns, which is considered a distinctive diagnostic feature (in about 83% of patients). [12]

The cause of this appearance may relate to the strands of fibrovascular stroma which present lower signal intensity in comparison with the remaining tumour on T2-weighted and post-contrast T1-weighted images. [13]

MR imaging shows the marked tendency of the tumour to displace rather than invade perineal structures, frequently crossing the pelvic diaphragm.

Extensive local excision reduces the risk of recurrence. Emerging therapies (hormonal therapy, embolisation and radiotherapy) minimise surgery and the treat of recurrences. [14-15]

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Aggressive perineal angiomyxoma, Myxoid leiomyoma, Myxoid lipomatous tumours, Infiltrating angiolipoma, Angiomyofibroblastoma

**Final Diagnosis:** Aggressive perineal angiomyxoma

**References:**


Description: T2-weighted fast spin-echo (FSE) imaging (a. and b. axial and c. coronal) shows a hyperintense perineal mass (star in a, b and c) with swirling appearance extending through pelvic diaphragm and displacing the urethra (arrow in a). Origin: "Department of Radiology. Hospital HM Torrelodones. Madrid"
Description: T2-weighted fast spin-echo (FSE) imaging (a. and b. axial and c. coronal) shows a hyperintense perineal mass (star in a, b and c) with swirling appearance extending through pelvic diaphragm and displacing the urethra (arrow in a). Origin: Department of Radiology. Hospital HM Torrelodones. Madrid
Description: T2-weighted fast spin-echo (FSE) imaging (a. and b. axial and c. coronal) shows a hyperintense perineal mass (star in a, b and c) with swirling appearance extending through pelvic diaphragm and displacing the urethra (arrow in a). Origin: Department of Radiology. Hospital HM Torrelodones. Madrid
**Description:** Axial T1-weighted fast spin-echo (FSE) imaging shows the isointensity of the mass (star) compared to the skeletal muscle. **Origin:** Department of Radiology. Hospital HM Torrelodones. Madrid
**Description:** Corresponding gadolinium-enhanced T1-weighted spin-echo with fat suppression imaging (a. and b. axial and c. coronal) shows a swirling pattern of heterogeneous enhancement within the tumour. Additional sagittal images (d and e) show the longitudinal extent of the mass. **Origin:** Department of Radiology. Hospital HM Torrelodones. Madrid
Description: Corresponding gadolinium-enhanced T1-weighted spin-echo with fat suppression imaging (a. and b. axial and c. coronal) shows a swirling pattern of heterogeneous enhancement within the tumour. Additional sagittal images (d and e) show the longitudinal extent of the mass. Origin: Department of Radiology. Hospital HM Torrelodones. Madrid
Description: Corresponding gadolinium-enhanced T1-weighted spin-echo with fat suppression imaging (a. and b. axial and c. coronal) shows a swirling pattern of heterogeneous enhancement within the tumour. Additional sagittal images (d and e) show the longitudinal extent of the mass. Origin: Department of Radiology. Hospital HM Torrelodones. Madrid.
**Description:** Corresponding gadolinium-enhanced T1-weighted spin-echo with fat suppression imaging (a. and b. axial and c. coronal) shows a swirling pattern of heterogeneous enhancement within the tumour. Additional sagittal images (d and e) show the longitudinal extent of the mass. **Origin:** Department of Radiology. Hospital HM Torrelodones. Madrid
Description: Corresponding gadolinium-enhanced T1-weighted spin-echo with fat suppression imaging (a. and b. axial and c. coronal) shows a swirling pattern of heterogeneous enhancement within the tumour. Additional sagittal images (d and e) show the longitudinal extent of the mass. Origin: Department of Radiology. Hospital HM Torrelodones. Madrid
**Figure 4**

**Description:** a) and b). Surgery confirmed a pedunculated partially encapsulated soft tissue mass extending through the pelvic diaphragm displacing adjacent structures without infiltration. **Origin:** Department of Gynecology Hospital HM Torrelodones. Madrid
Description: a) and b). Surgery confirmed a pedunculated partially encapsulated soft tissue mass extending through the pelvic diaphragm displacing adjacent structures without infiltration. Origin: Department of Gynecology Hospital HM Torrelodones. Madrid