

Vulvar leiomyosarcoma in Bartholin's gland

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Section: Genital (female) imaging

Area of Interest: Pelvis

Procedure: Diagnostic procedure

Imaging Technique: MR

Special Focus: Neoplasia Case Type: Clinical Cases

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Patient: 50 years, female

Clinical History:

The patient presented with a right labia majora nodule. She was diagnosed with infected Bartholin's cyst and started antibiotic therapy. Nevertheless, the lesion showed progressive enlargement during the following month. Drainage of the mass followed, complicated with lesion haemorrhage. Pelvic MRI was performed to help with diagnosis and treatment planning.

Imaging Findings:

MRI depicted a heterogeneous mass, mainly of intermediate signal intensity on both T1 and T2-weighted images (Fig. 1, 2) and areas of high-signal-intensity on T2-weighted sequences, corresponding to cystic necrosis on pathology (Fig. 2). The lesion showed heterogeneous contrast enhancement, with early peripheral enhancement and progressive fill-in (Fig. 3). Restricted diffusion was demonstrated within the mass on diffusion-weighted images (Fig. 4).

Surgery followed and the lesion was excised. Histopathologic examination revealed a vulvar leiomyosarcoma, of more than 5 cm in diameter, infiltrating margins, cell atypia and a proliferative index of 20%. CT body scan was negative for the presence of metastatic lymphadenopathy and distant metastases.

Discussion:

Even if soft tissue tumours are very rare, leiomyosarcoma is the most common sarcoma of the vulva, representing 1% of vulvar malignancies [1, 2, 4]. It originates from the smooth muscle containing erector-pili muscle, blood vessels walls [1] and round muscle or dartos muscle of the recto-vaginal septum [4, 5]. The most frequent location is the upper-vagina [3] and it usually occurs in women between 40 and 50 years [1]. Radiation therapy in the genital tract has been reported as a risk factor [3].

The most common clinical presentation is a painless and rapidly-growing submucosal mass in the labia majora [1, 3, 4, 5]. Other symptoms are pain, pruritus or erythema [1]. Any vulvar lump with unusual characteristics such as rapid growth or hard consistence in Bartholin's gland area should be investigated carefully to avoid delay in diagnosis [1, 2, 4, 5].

Diagnosis is usually based on histopathological examination of excised lesions taking into account that cytology may suggest benignity. It requires at least 3 of the following characteristics: atypical cells, mostly large and hyper-

chromatic cells forming whirling and bundles, high proliferation index, infiltrating margins and more than 5 cm in diameter [2, 4, 5].

MRI is the modality of choice to allow proper tumoral delineation, evaluation of the local extent of the disease and its relationship to adjacent structures, to aid in surgical planning and to reduce surgical morbidity. Imaging findings of vulvar leiomyosarcomas are variable, such as of leiomyosarcomas in other locations. The tumour is usually described as a heterogeneous and irregularly-shaped mass with areas of high T2 signal due to cystic necrosis, and areas of high T1 signal due to haemorrhage. It often depicts heterogeneous contrast enhancement and areas of restricted diffusion on diffusion-weighted imaging. High signal intensity is detected on STIR sequences, without areas of fat [3].

CT body scanning is necessary to exclude lymphatic and haematogenous spread of the tumor. However, leiomyosarcomas localized in the Bartholin's gland area are often less aggressive [1, 4].

Currently, surgery is the treatment of choice. The role of radiotherapy and chemotherapy remains uncertain [1, 2, 4].

Differential Diagnosis List: Bartholin's gland area leiomyosarcoma, Squamous cell carcinoma, Adenocarcinoma, Melanoma, Spindle cell synovial carcinoma, Paraganglioma, Bartholin's gland abscess, Bartholin's gland cyst

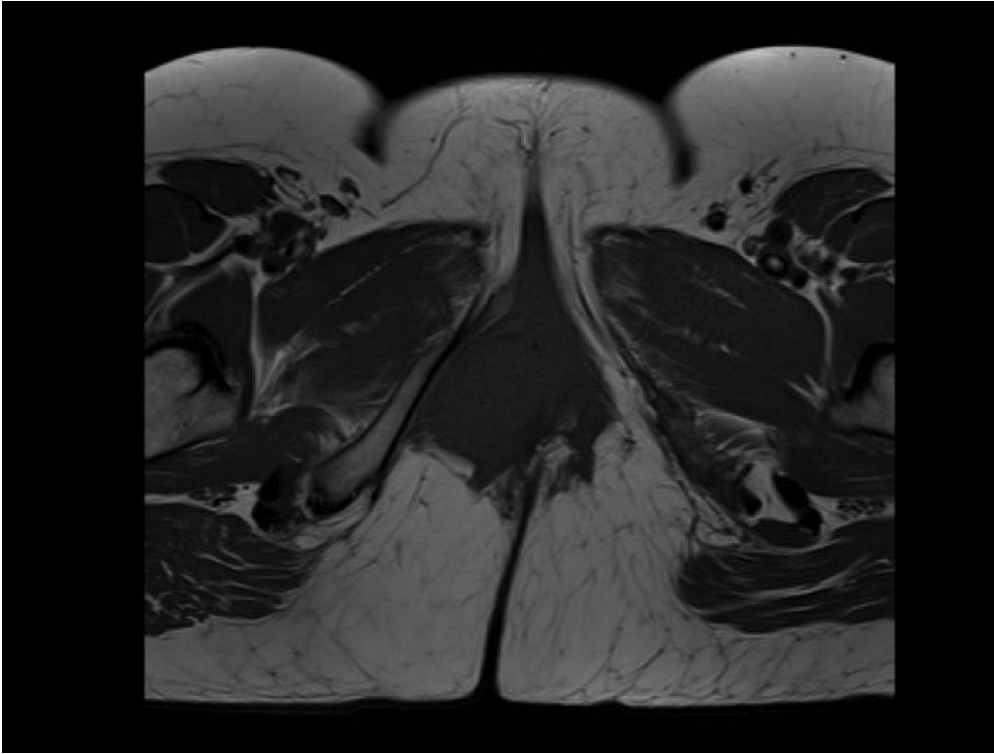
Final Diagnosis: Bartholin's gland area leiomyosarcoma

References:

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Figure 1

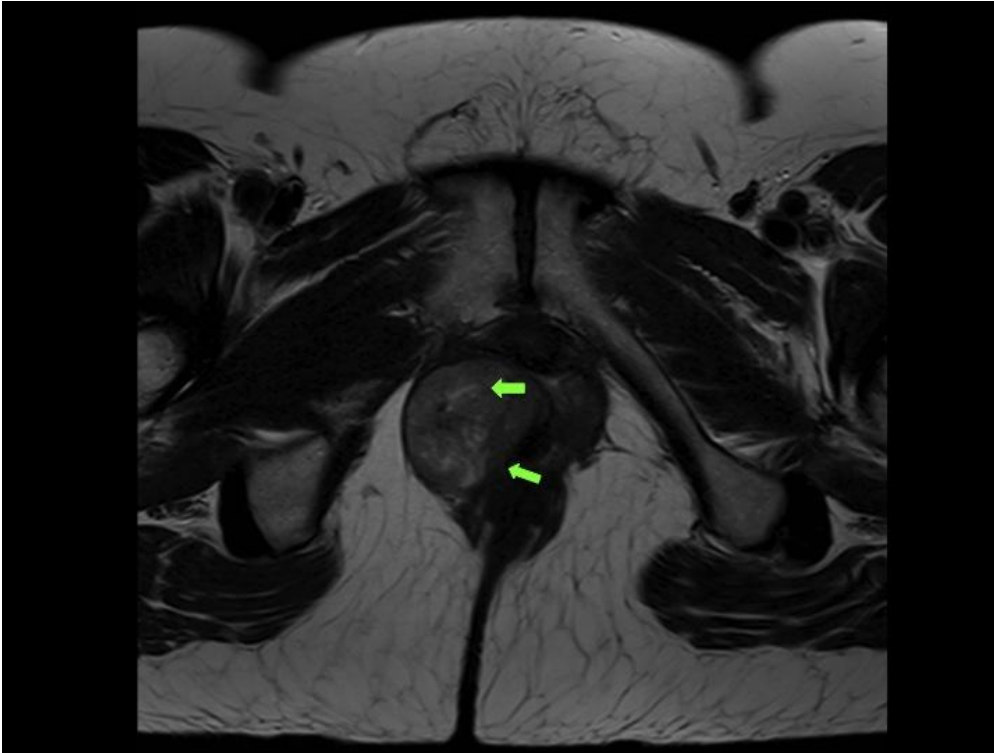
a



Description: Right vulvar mass of intermediate signal intensity. **Origin:** Hospital Universitario Basurto

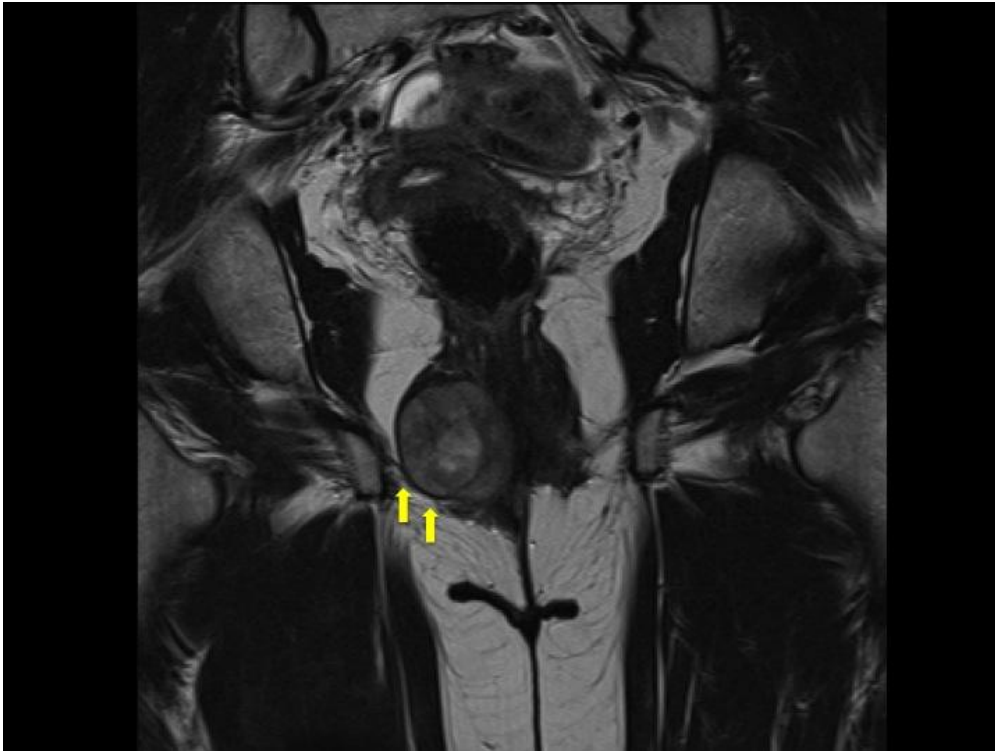
Figure 2

a



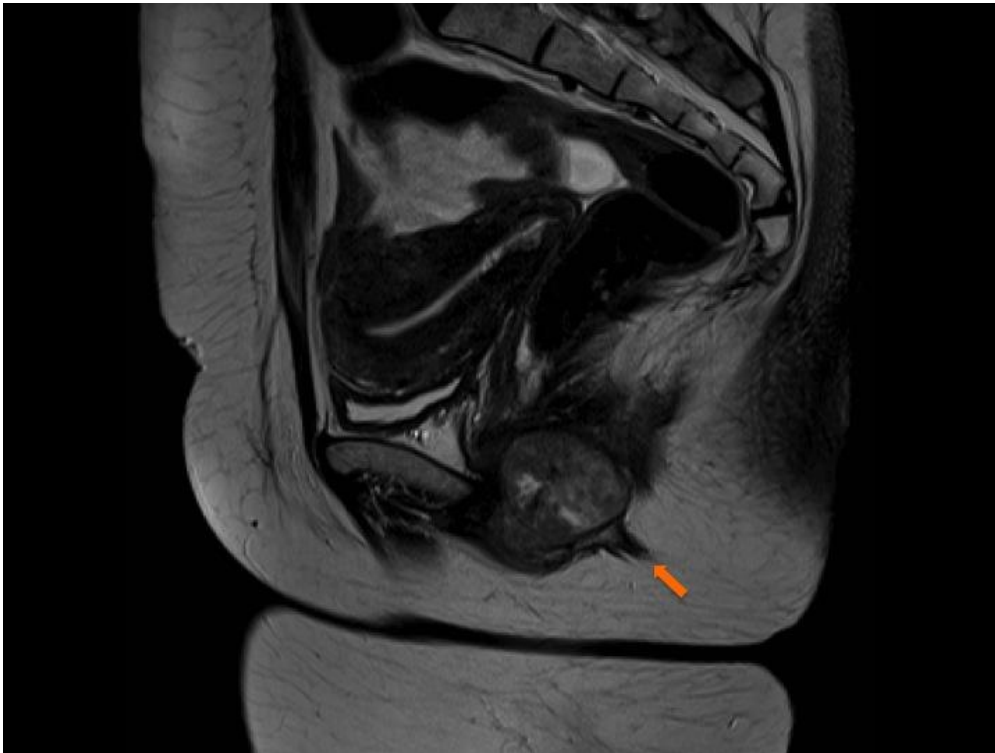
Description: Axial T2 TSE. Relative isointense mass centred in the right vulva, replacing the labia majora and showing central areas of hyperintensity (green arrows). **Origin:** Hospital Universitario Basurto

b



Description: Coronal T2 TSE. The mass is located in Bartholin's gland area and extends to the right levator muscle of anus (yellow arrows), without showing invasion. **Origin:** Hospital Universitario Basurto

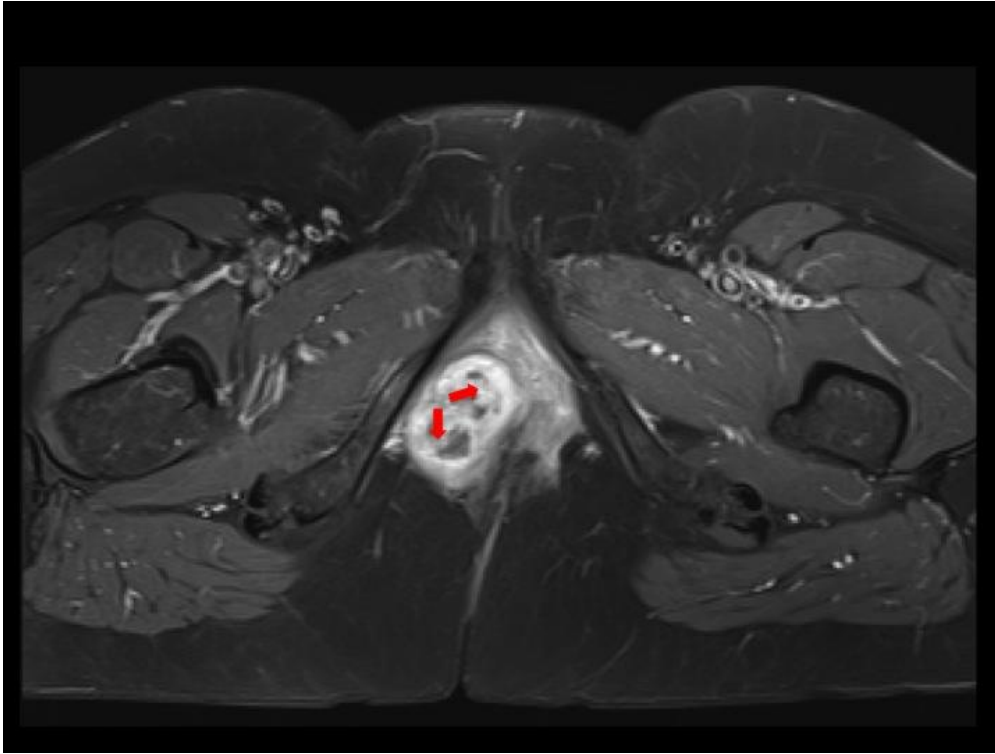
c



Description: Sagittal T2 TSE. The lesion is superior to the perineal membrane (orange arrow). **Origin:** Hospital Universitario Basurto

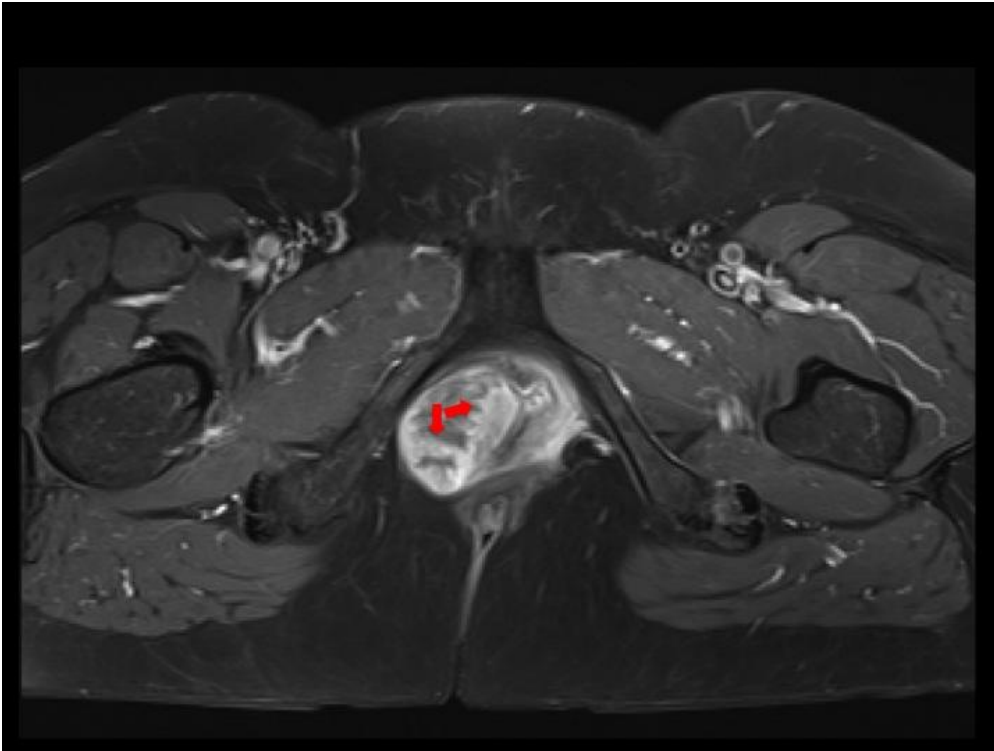
Figure 3

a



Description: Early phase: the mass depicts heterogeneous, mainly peripheral enhancement. **Origin:** Hospital Universitario Basurto

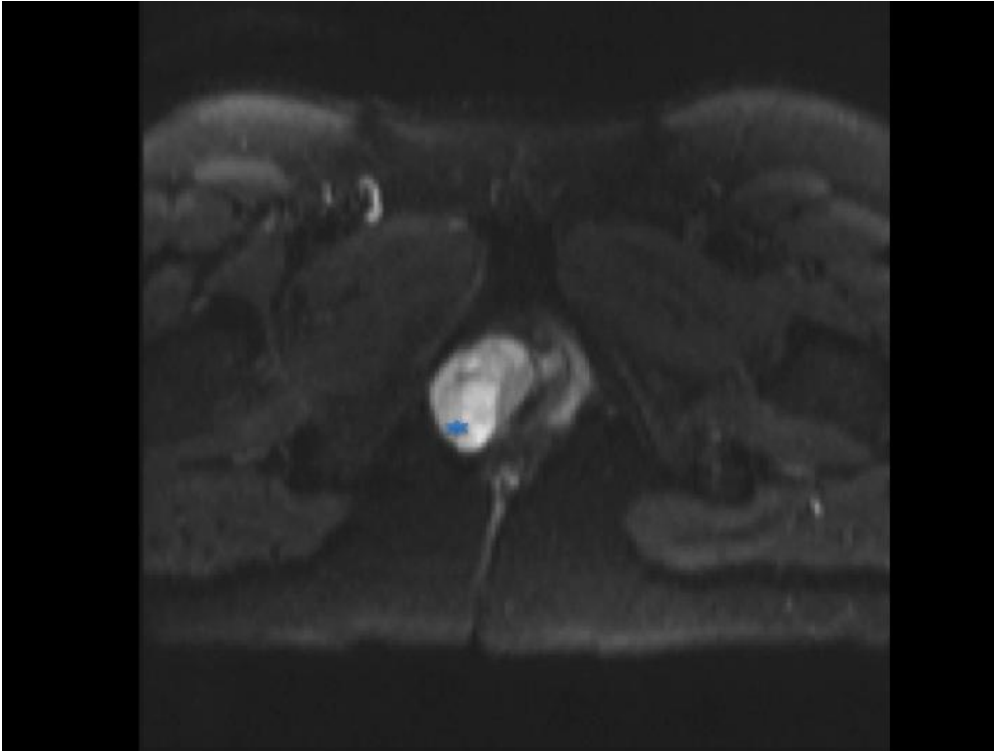
b



Description: Delayed phase: progressive and heterogeneous fill-in of the lesion. **Origin:** Hospital Universitario Basurto

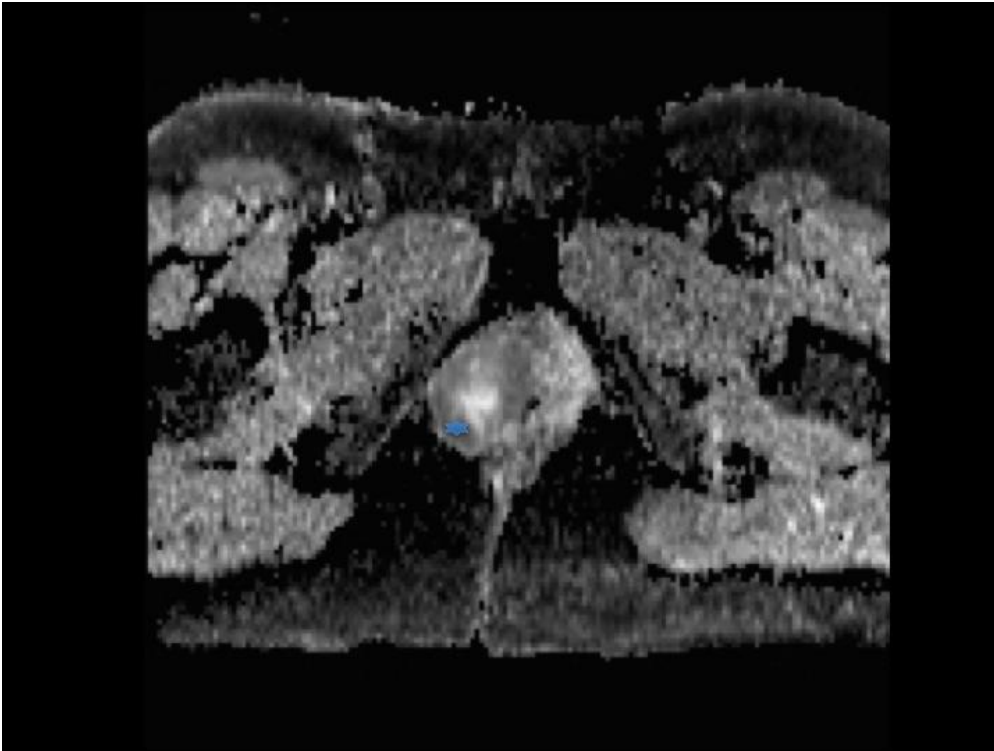
Figure 4

a



Description: Heterogeneous, high-signal-intensity mass (asterisk) on high b-value DWI. **Origin:** Hospital Universitario Basurto

b



Description: Heterogeneous, low-signal-intensity mass (asterisk), confirming the restricted diffusion on ADC map. **Origin:** Hospital Universitario Basurto