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Case report

Perineal and pelvic aggressive angiomyxoma: Imaging finding in an uncommon case report ☆☆☆

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ABSTRACT

Aggressive angiomyxoma is a rare benign and locally invasive mesenchymal tumor that is found most frequently in women at reproductive age. We report typical CT and MRI appearances of a 36-year old young woman with an aggressive angiomyxoma of the pelvis and perineum that was proved by ultrasound guided biopsy. This study describes the imaging features of these tumors, which may help to approach the diagnosis by their distinctive imaging with high signal intensity on T2-weighted image related to the myxomatous stroma and their characteristic of swirling or layering internal pattern after intravenous gadolinium contrast administration. We also review the CT and MRI features of this disease in the current literature.

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Introduction

Aggressive angiomyxoma is a rare, benign, locally infiltrative mesenchymal tumor found usually in young women. It was first described by Steeper and Rosai in 1983 [1].

It's a slow-growing and low-grade neoplasm involving the pelvis and perineum occurring mostly in women of reproductive age, with a high risk for local recurrence. However, there were also some cases occurring in the scrotum or spermatic cord in males [2].

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Fig. 1 – Clinical appearances of the tumor. A 36-year-old woman, with a large pelvic and perineal mass, enlarging in size within 02 years, mimicking a massive perineal non reducible hernia without other abnormalities.

The tumor's localization makes it difficult to distinguish it from others entities such as Bartholin's gland cyst, vaginal cyst, abscess, leiomyoma, lipoma or hernia [9]. In all cases, the diagnosis of certainty can only be proved by histology [3].

Both CT and MR imaging plays a key role in making an accurate diagnosis and determining the extent of these tumors [4].

Case presentation

A 36-year-old woman, without history of illness or drug use, presented to our radiology department with a large pelvic and perineal mass, evolving for 2 years ago, and increasing progressively in size.

The clinical examination revealed a massive perineal non reducible hernia without other abnormalities (Fig 1). Biologically, inflammatory tests were negative.

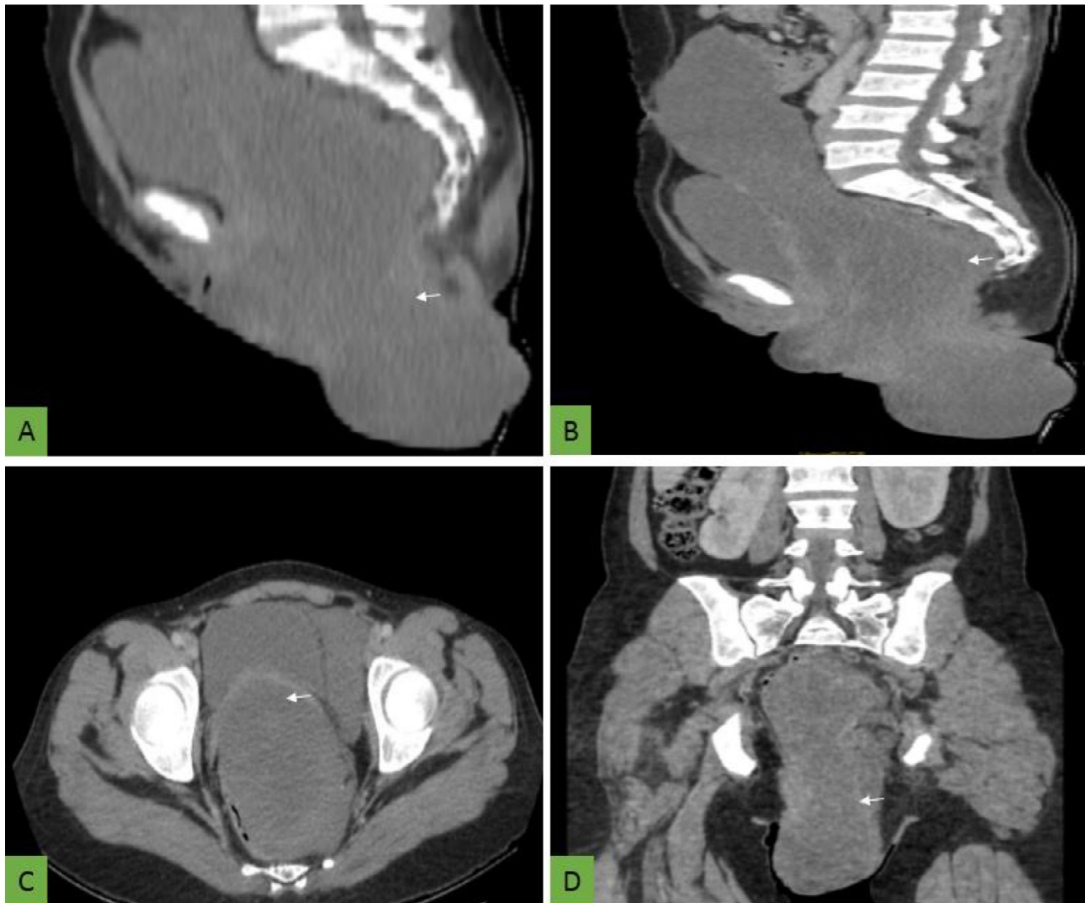


Fig. 2 – CT appearances of the tumor (A) sagittal unenhanced CT scan shows a pelvic mass displacing bladder, uterus, and distal rectum, with attenuation less than that of muscle. this mass traverses the pelvic diaphragm sagittal (B), axial (C), and coronal (D) enhanced CT scan reveals a low enhancement according to a swirling model within the tumor, more marked on the sagittal section (B).

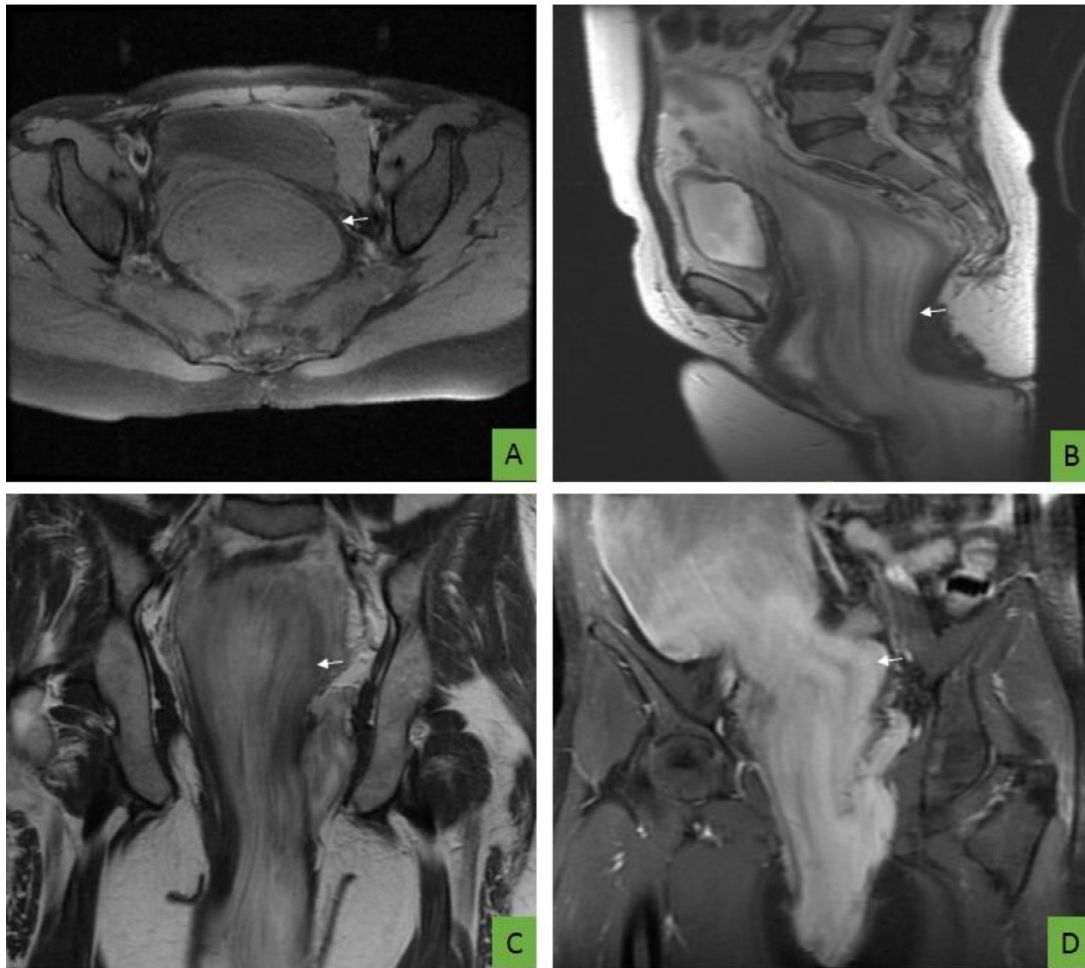


Fig. 3 – MRI appearances of the tumor (A) axial unenhanced t1-weighted MR imaging with fat suppression shows a pelvic mass is primarily isointense to muscle (arrow) (B, C) sagittal and coronal t2-weighted MR imaging displays the tumor with high signal intensity interspersed with swirled or layered strands of lower signal intensity (arrow) (D) coronal enhanced t1-weighted MR imaging with fat suppression reveals the swirling (or layering) pattern of remarkable enhancement within the tumor (arrow) and the tumor traverses the pelvic diaphragm.

CT and MRI scans were performed to evaluate the mass. Unenhanced CT scan showed a large mass displacing the bladder, uterus, and rectum, with an attenuation less than that of muscle. This mass crosses the pelvic diaphragm and is exteriorized at the perineum. (Figs. 2A, B, and C). On contrast-enhanced CT, the mass revealed mild enhancement with an internal swirling pattern of enhancement within the tumor more marked on the sagittal enhanced CT (Figs. 2D, E, and F).

On MR imaging, the mass was primarily isointense compared to the muscle on T1-weighted images with fat suppression. (Fig. 3A). On T2-weighted images, high signal intensity of the mass interspersed with swirled or layered strands of lower signal intensity was seen (Figs. 3B, C, D, and E). The mass was markedly enhanced in a swirling (or layering) manner on the coronal contrast-enhanced T1-weighted MR imaging with fat

suppression (Fig. 3F). The final diagnosis was proved by ultrasound guide biopsy.

Histopathological sections of the tumor showed many walled vessels of various sizes, a loose myxoid and collagenous stroma and stellate and spindle-shaped neoplastic cells. A tumor cell usually possessed 1 nucleus and showed neither apparent nuclear atypia nor mitotic figures (Fig. 4).

The positive immunohistochemistry results of strong positivity for AML (Fig. 5A), desmin (Fig. 5B) and moderate positivity for CD34 (Fig. 5C). The tumor cells showed positive immunoreactivities for estrogen and progesterone hormone receptors (Figs. 5D and E).

A neoadjuvant hormonal therapy with analogues of Gonadotrophin-releasing hormone analogues (Decapeptyl) was introduced to reduce the tumor size in order to optimize the surgical results.

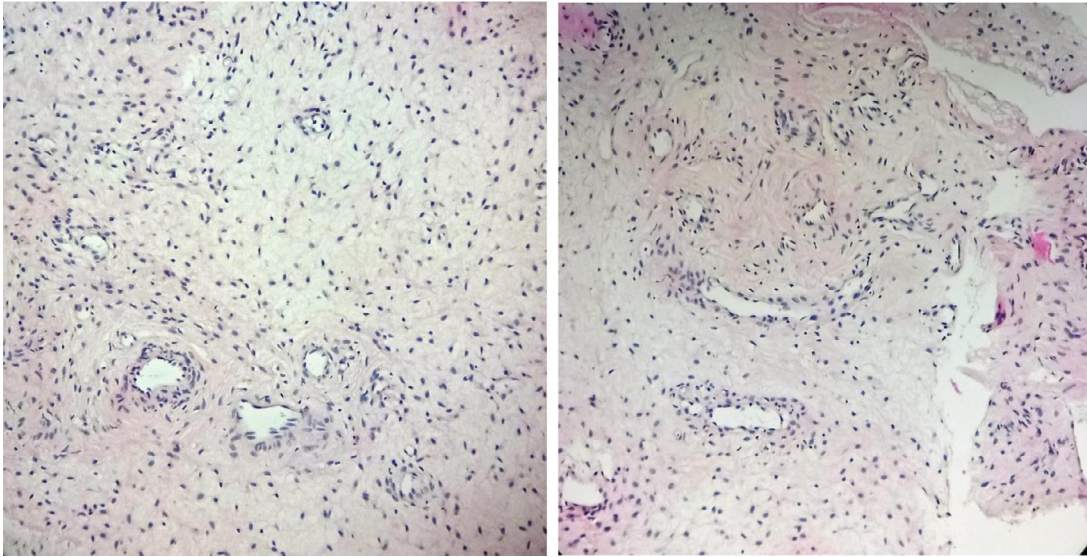


Fig. 4 – Histopathological appearances of the tumor histopathological study showed a tumor proliferation on a myxoid and fibrous background, with small rounded or elongated cells with stellate outlines and eosinophilic cytoplasm. they are no cytonuclear atypia or mitosis.

Discussion

Aggressive angiomyxoma is a benign mesenchymal tumor with a predilection for involving the perineum and pelvis of pre-menopausal women, usually between the second and fourth decade [5]. It's still called aggressive because of the high rate of the local recurrence after resection [6].

Clinically, these tumors are usually polypoid and most often confined to the vulva, lower pelvis and perineum. They are mostly asymptomatic, but they may present with symptoms and signs suggestive of an obturator and perineal hernia [7].

In imaging, it differs from a diffuse peritoneal leiomyomatosis due to its extraperitoneal localization, and also from a rectal GIST due to fatty interface with the digestive wall, between the lesion and the rectal muscular wall [8].

CT or MRI imaging plays an important role in approaching the diagnosis and establishing the optimal surgical approach [4]. In any case, the certain diagnosis can only be proved by histology [4].

On CT, it is of low attenuation compared to the muscles. On MRI, these tumors are isointense to the muscle on the T1 weighted image, hyperintense on T2-weighted image and they show an enhancement after intravenous gadolinium injection with a characteristic of swirling or layering internal pattern [9].

The low attenuation on CT and their high signal intensity on MRI are likely to be related to its high water content and to the loose myxoid matrix of angiomyxoma [9].

On MRI findings, the swirling or layered strands are of slightly lower signal intensity than the rest of the tumor on T2-weighted MR images, and are also evident on contrast-enhanced T1-weighted MR imaging with fat suppression [9].

These tumors, generally have a well-defined margin, displacing rather than invading adjacent organs, such as urethra, vagina, anus the sphincter and the rectum [10].

In addition to the precise diagnosis of the tumor, preoperative imaging is very useful to assess the extent of the tumor and helps to determine the surgical approach.

Histologically, these lesions contain connective cells dispersed on a background of myxoid and collagen with rich vascularization [11]. There is no cytonuclear atypia or mitosis. The tumor cells express the hormone receptors for estrogen and progesterone, thus confirming the hormone-dependent nature of these tumors [11].

Treatment of these tumors is always surgery, but their risk of recurrence is high. Only radical surgery with good clear excision margin on histology associated to hormonal treatment using Gn-RH analogues can avoid the high risk of recurrence [10].

Given the hormone-dependent nature of these tumors, adjuvant and neo adjuvant hormonal therapy such as GnRh agonists, with long-term follow-up was considered as a potential therapeutic target to reduce the tumor volume and optimize the results of surgical treatment [11].

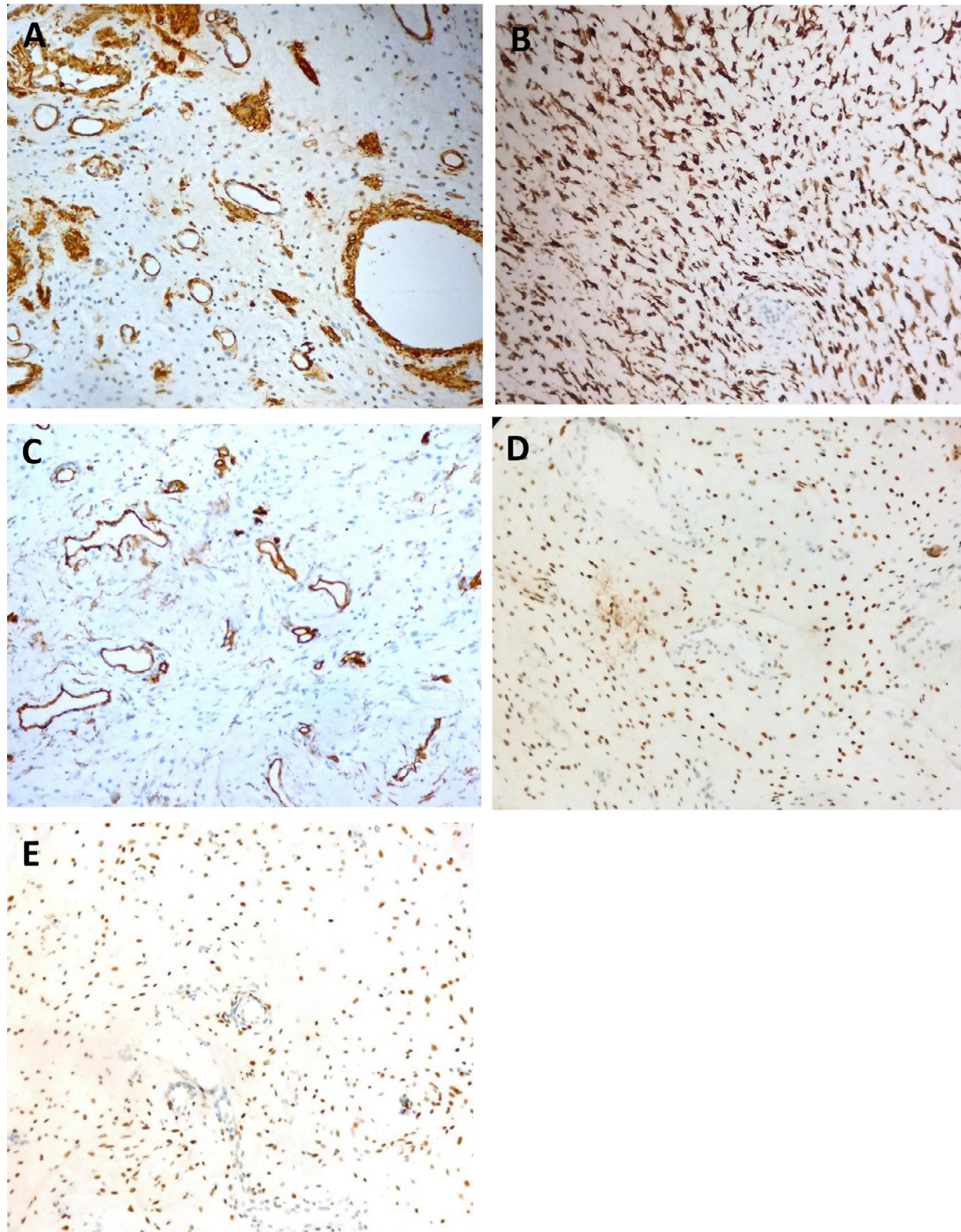


Fig. 5 – Immunohistochemical studies of the tumor. Most of the tumor cells are immunopositive for AML (A), DESMINE (B), and hormonal receptors RP (D), RO (E). The tumor cells are occasionally immunopositive for CD34 (C).

Conclusion

In summary, angiomyxoma is a rare, locally aggressive mesenchymal tumor, which can easily be misdiagnosed with other diseases just like Bartholin's gland cyst.

We found that MRI and CT imaging can be useful to approach the diagnosis of these tumors by their distinctive

imaging appearance of swirled or layered internal architectures after enhancement.

The key role of imaging is to specify the extent of these tumors and to determine the optimal surgical approach.

In any case, the diagnosis of certainty is histological

Only radical surgery with long term follow up may help to avoid a recurrence.

Ethics approval and consent to participate

Oral and signed consent was obtained from the patient concerned. The study was conducted anonymously.

Availability of data and materials

The data sets are generated on the data system of the CHU Hassan II of Fes, including the data of the anatomopathological analysis.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2021.04.041](https://doi.org/10.1016/j.radcr.2021.04.041).

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