Radiographic diagnosis and differentiation of an aggressive angiomyxoma in a male patient

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ABSTRACT

Aggressive angiomyxoma is a rare soft-tissue tumor which usually occurs in female patients of reproductive age. Its occurrence in men is even more unusual and as illustrated in this case the difference between pathology suggested by a physical examination and its actual extent can be quite striking. We present a case report of an 81-year-old man with the typical MRI appearances of a pelvic aggressive angiomyxoma, describe imaging and histopathologic features of this rarely seen locally infiltrative neoplasm and also discuss therapeutic options for patients with an aggressive angiomyxoma.

CASE REPORT

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An 81-year-old man presented to the surgery outpatient clinic with a swelling of the right perineum that was slowly growing for about one year. His complaints involved defecation problems and a feeling of discomfort during prolonged seating. Physical examination demonstrated an indolent, soft lump of the perineal region, adjacent to the posterior midline, about 4 x 5 cm big, with ill-defined edges. Apart from slightly reddened skin in this location (attributed to constant pressure application and irritation from seating) no other apparent visual changes were noted. Bloodcounts were unremarkable. MRI examination was performed.

IMAGING FINDINGS

MRI revealed a complex, hourglass-shaped, pelvic soft tissue mass with a 15 cm cranio-caudal and a 5 x 7.5 cm axial extension (Figure 1). The tumor was hyperintense to muscle on T2-weighted (Figure 1) and isointense on T1-weighted images (Figure 2). After administration of intravenous contrast medium the mass showed a strong enhancement with swirled strands of hypointense tissue in the mass center (Figure 3). The distal component of the tumor was divided into several parts. The largest part extended dorsally into the right perineal region (Figure 4) and was situated under the gluteus maximus muscle. Smaller anterior parts, which diverged at the proximal aspect of the penile corpus spongiosum, extended bilaterally into the perineal area passing along the crus of the corpora cavernosa (Figure 4). The lesion was rather well circumscribed and did not show a frankly infiltrative pattern into the surrounding soft tissues.

MANAGEMENT

Core needle biopsy did not yield enough material for definite pathological assessment. Thus an open incisional biopsy was performed. The biopsy specimen showed a solid mass of whitish-grey color with a fibrous appearing surface after sectioning. Histologically, the tumor consisted of a highly vascular loose fibrotic stroma in which relatively small and uniform spindle cells were embedded. The cellularity was moderate and the spindle cells showed monomorphic nuclei with distinct nucleoli. Mitoses were nearly completely absent

(< 1 / 10 HPF). The vessels showed a mixture of smaller thin-walled capillaries and larger vascular channels with prominent muscular walls (Figure 5-6). Immunohistochemically, the tumor cells showed an expression of desmin, smooth muscle actin, CD34, estrogen receptor, and progesteron receptor. The diagnosis of aggressive angiomyxoma (AAM) was made.

Because of the complaints of the patient, we planned a resection of the extrapelvic part of the disease solely to relieve the problems with sitting. The infralevator, right-sided part of the tumor was removed with an uneventful postoperative course. The patient was put on antihormonal treatment with tamoxifen (40 mg daily) after surgery.

FOLLOW-UP

The latest follow-up 16 months following R2 resection showed a stable disease without signs of progression. The patient still had signs of obstipation but was otherwise asymptomatic.

DISCUSSION

ETIOLOGY & DEMOGRAPHICS

Aggressive angiomyxoma is a rare (about 350 known cases) soft tissue tumor which occurs predominantly in premenopausal women (male to female ratio = 1:6). It usually involves the pelvis, in particular the perineum, and is characterized by a locally aggressive growth pattern. There is generally no involvement of adjacent organs which are often displaced by the tumor. Consequently, the tumor can already have a considerable size at the time of diagnosis. AAM is known to recur in about one third of cases if not completely excised, which can prove to be technically challenging [1].

AAM was first described in 1983 by Steeper and Rosai in a female patient [2]. Bégin et al. reported an AAM in a male patient about two years later [3]. To our knowledge, fewer than 50 cases have been reported in males up to now and imaging reports are scarce [4-10].

CLINICAL & IMAGING FINDINGS

The published cases involved perineum, scrotum, testis, epididymis, inguinal area, and spermatic cords and have been partly mistaken for hernia, hydrocele and spermatocele on initial physical exams [4]. The most common manifestation is a slowly growing soft tissue mass with diameters ranging from 2 to 36 cm [4]. The average age of male patients with AAM is 46 years but rare cases in infants and older patients (81 years) have been described [4, 11].

MRI is the best modality for AAM imaging because of its ability to visualize a transdiaphragmatic spread and the relationship of the mass with the pelvic organs, which is crucial for surgical planning. AAM is isointense to muscle on T1-weighted images and hyperintense on T2-weighted sequences owing to its loose and sometimes myxoid stroma. The tumor may present as a cystic lesion on ultrasound, which may lead to misdiagnosis of the lesion as simple Bartholin's or vaginal cysts in female patients [12]. Typical MRI features of AAM are swirled strands aligned with the craniocaudal axis [13, 14]. Outwater at al. suggested that this particular imaging feature is caused by stretching of the fibrovascular stroma during protrusion through the pelvic diaphragm [8]. After administration of i.v. contrast medium a strong enhancement is observed. Histologically, AAM is generally composed of a fibromyxoid stroma with widely scattered spindled to stellate-shaped cells and variably sized, thin- and thick-walled vessels. Mitotic activity is very low and atypical mitoses are absent. Immunohistochemically, the spindle cells of AAM typically show positivity for desmin, CD34 and smooth muscle actin. Additionally, estrogen and progesterone receptors are consistently expressed in these tumors, suggesting a hormonal role in tumor development [15].

TREATMENT & PROGNOSIS

Because of its locally infiltrative growth and the high risk of local recurrence (in up to 72% of cases) the treatment of choice is an excision with wide margins. Less radical surgery with an adjuvant therapy including radiotherapy, chemotherapy, vessel embolization, and hormone suppression has also been discussed in the literature [1, 8, 16, 21].

Since recurrences have been described to generally have a similar appearance like the primary tumor, the most useful follow-up imaging modality seems to be MRI. AAM is usually a nonmetastasizing tumor but two cases of pulmonary and mediastinal involvement have been described [17].

DIFFERENTIAL DIAGNOSES

The differential diagnoses in male patients include: angiomyofibroblastoma, myxoid neurofibroma and pelvic myxoma.

Angiomyofibroblastoma presents on MRI as a well circumscribed mass that is isointense to muscles on T1-weighted and hyperintense on T2-weighted images owning to its high content of loose edematous connective tissue. After administration of gadolinium containing contrast media the tumor shows a strong homogenous enhancement [18].

T2-weighted MR images of myxoid fibroma show a well-defined round tumor in the pelvic cavity with mixed low and high signal intensity due to myxoid degeneration. After contrast administration the tumor displays a heterogenous enhancement. In some cases the presentation on T2-weighted images is that of a targetlike lesion with high signal in the periphery (myxoid stroma) and hypointense center (fibrous-collagenous tissue) [19].

Pelvic myxomas are well circumscribed tumors, homogenously hypointense on T1w and hyperintense on T2w images. On contrast enhanced MR imaging they present heterogeneous signal intensity with nonenhancing cystic areas [20].

In summary, AAM is a rare tumor, with typical imaging features on MR imaging - a pelvic soft-tissue mass of high signal intensity interspersed with longitudinal strands of lower signal intensity on T2-weighted images and heterogeneous enhancement after contrast administration. A translevator spread and displacement of adjacent pelvic organs are often present at the time of diagnosis.

TEACHING POINT

Aggressive angiomyxoma typically presents as an hourglass-shaped, pelvic soft tissue mass that is hyperintense to muscle on T2-weighted and isointense on T1-weighted images. After administration of intravenous contrast medium it shows a strong enhancement with swirled central strands of hypointense tissue.

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FIGURES

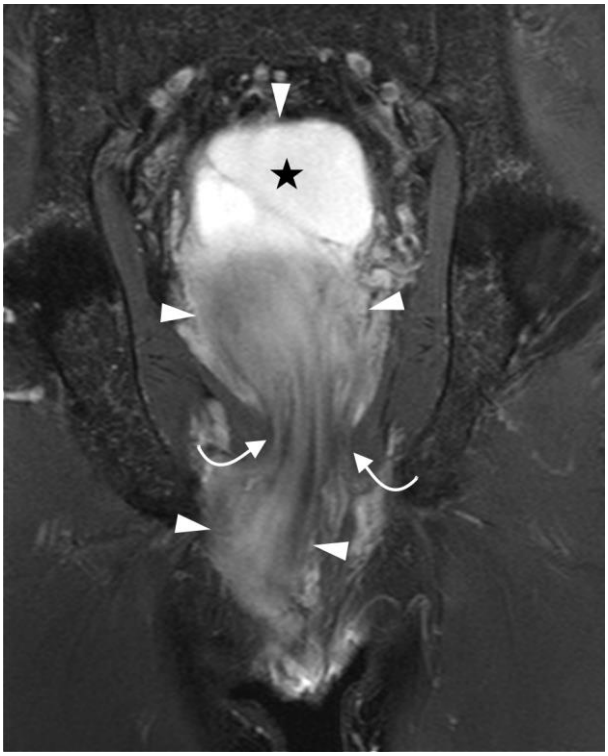

Figure 1: 81-year-old male with an aggressive angiomyxoma. FINDINGS: heterogeneous, hyperintense pelvic mass (white arrowheads, 15 x 5 cm) with a cystic component at the upper aspect (black star). The mass extends both superiorly and inferiorly relative to the levator ani muscle (curved white arrows). The internal architecture is composed of hypointense layered strands. TECHNIQUE: Coronal fast spin-echo T2-weighted fat-saturated MR image (repetition time msec/echo time msec, 4472/60, 3 Tesla)

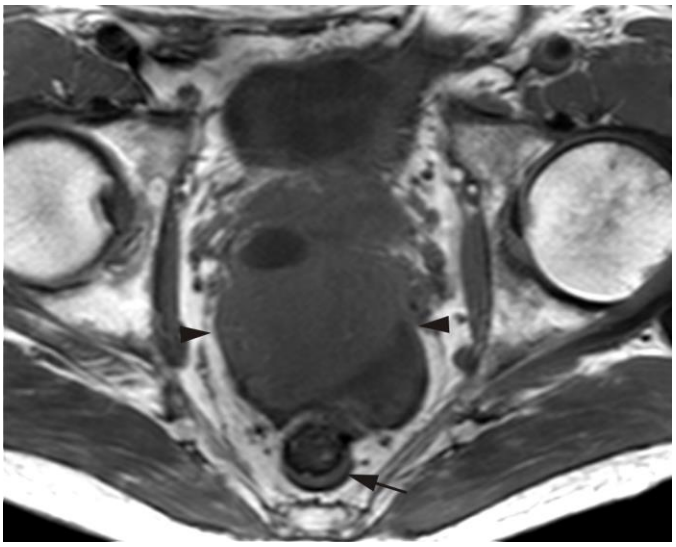

Figure 2: 81-year-old male with an aggressive angiomyxoma. FINDINGS: Well-defined mass (black arrowheads, 5 x 7,5 cm) situated centrally in the pelvis and anteriorly to the rectum (black arrow). The signal intensity is isointense to that of muscle. TECHNIQUE: Axial T1-weighted spin-echo (713/9, 3 Tesla) MR image

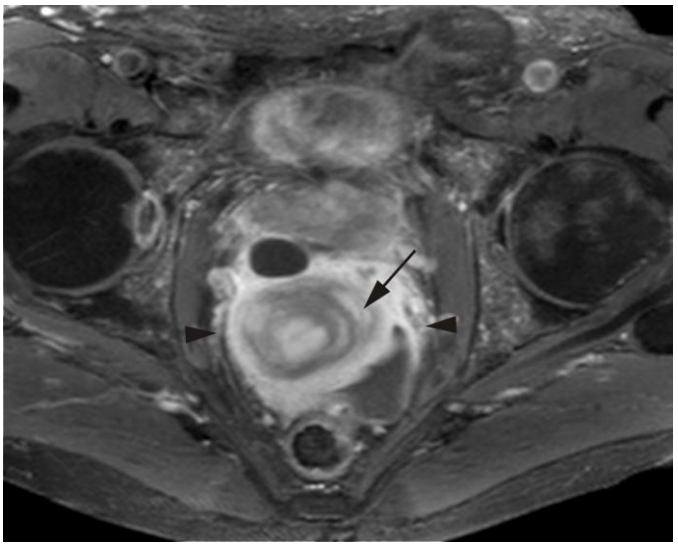

Figure 3: 81-year-old male with an aggressive angiomyxoma. FINDINGS: Image obtained at the same level as in Figure 2 demonstrates a heterogeneously enhancing mass (black arrowheads, 7,5 x 5 cm). Swirled strands of tissue (black arrow) appear less enhanced in the mass center. TECHNIQUE: Axial fat-supressed T1-weighted spin-echo (666/9, 3 Tesla) delayed post-contrast MR

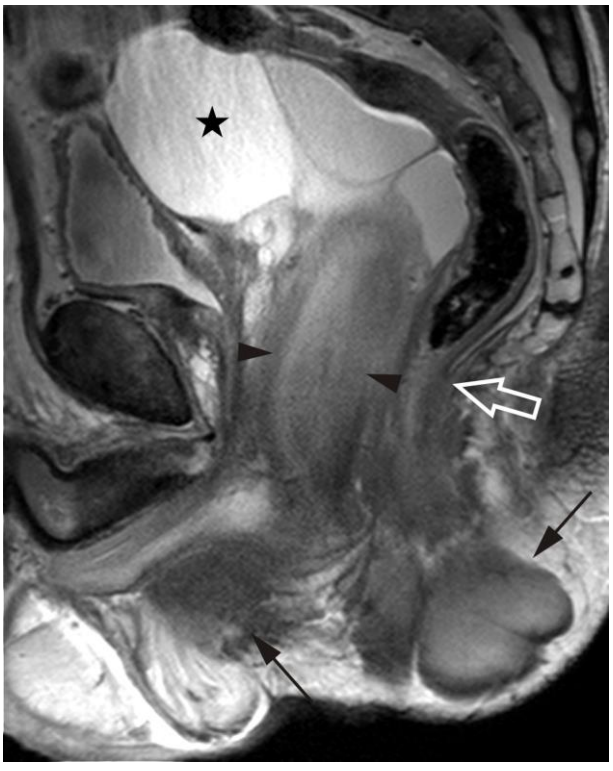

Figure 4: 81-year-old male with an aggressive angiomyxoma. FINDINGS: Image demonstrates the multidirectional extension of the mass (black arrowheads) relative to the pelvic diaphragm. The mass lies anteriorly to the rectum (open white arrow) and involves the perineum dorsally and anteriorly (black arrows, 4 x 3,5 cm and 4 x 4 cm respectively). Cystic changes can be seen in the upper part of the mass (black star, 4,5 x 5,5 cm). TECHNIQUE: Sagittal fast spin-echo T2-weighted MR image (4721/90, 3 Tesla)

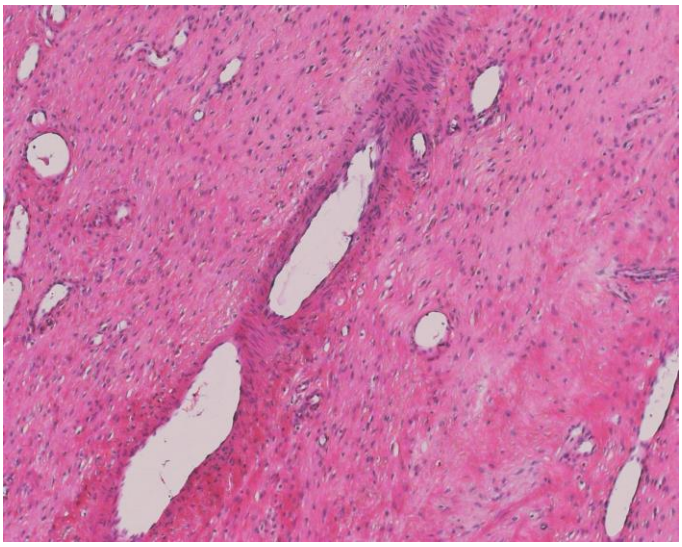

Figure 5: 81-year-old male with an aggressive angiomyxoma. Histologically, the tumor shows a monotonous fibrous stroma with bland spindle cells and abundant vascular channels. The centrally located vessels demonstrate a prominent muscular wall (HE, 200x)

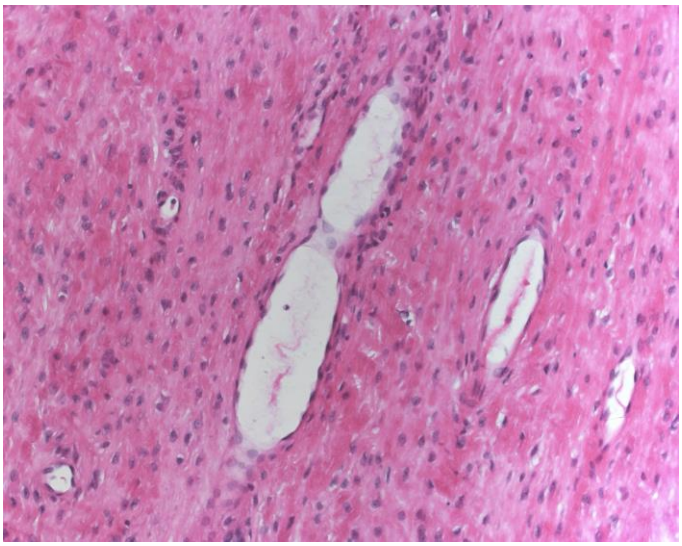

Figure 6: 81-year-old male with an aggressive angiomyxoma. In greater magnification the spindle cells show monomorphic nuclei with prominent nucleoli. Mitotic figures are absent. The vessels shown here lack a muscular wall and have a capillary phenotype (HE, 400x)

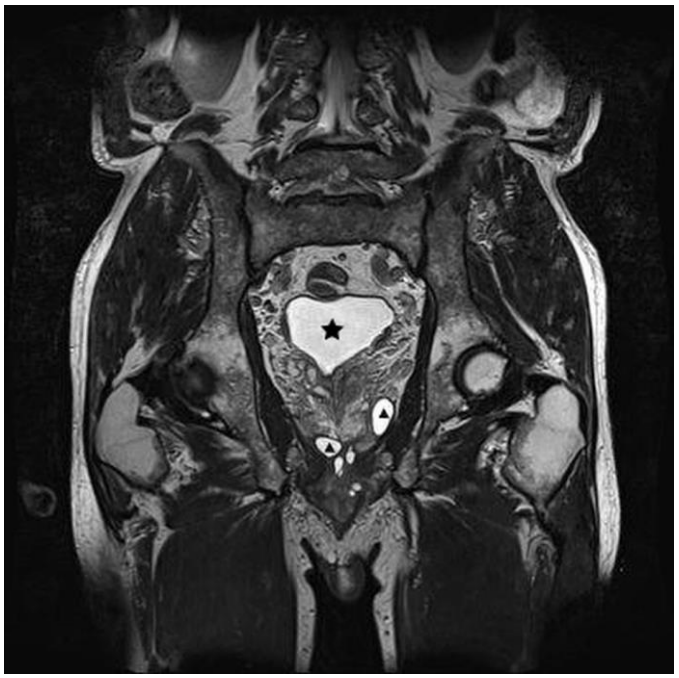

Figure 7: 81-year-old male with an aggressive angiomyxoma. FINDINGS: About 2 years after R2 resection imaging shows stable findings - some residual cystic lesions are still present (black triangle, ca, 2-3 cm in diameter) but the main tumor mass has been removed and shows no signs of recurrence. The urinary bladder is marked by a black star. TECHNIQUE: Coronal fast spin-echo T2-weighted fat-saturated MR image (1600/100, 3 Tesla).

Etiology	No etiologic factors are known.
Incidence	About 350 known cases.
Gender ratio	Male to female ratio = 1:6
Age predilection	Premenopausal women
Risk factors	Premenopausal women
Treatment	• Radical surgery with wide margins. • Surgery with chemotherapy and radiotherapy. • Hormonal therapy (GnRH agonists)
Prognosis	• The prognosis is very good (only 2 cases with fatal metastatic disease reported). Common recurrences (9 to 72 %)
Findings on imaging	• On MRI pelvic hourglass-shaped soft tissue mass, hyperintense to muscle on T2-weighted and isointense on T1-weighted images. After intravenous contrast medium strong enhancement with swirled central strands of hypointense tissue.

Table 1: Summary table for aggressive angiomyxoma

Pathology	MR	CT	Ultrasound
Angiomyo-fibroblastoma	• Well circumscribed • Low signal intensity on T1w- images • High signal intensity on T2w- images • Strong homogenous enhancement	• Inhomogeneous hypodense mass with heterogeneous enhancement. [11]	• Mixed echoic mass with cystic changes.
Myxoid neurofibroma	• Heterogeneously hyperintense on T2w images • Heterogeneous enhancement • Sometimes target-like appearance: T2w hyperintense periphery, T2w hypointense center.	• Low-attenuating cystic components depending of the proportion of myxoid degeneration. • Occasionally target-like enhancement pattern similar as MRI. [19]	• Occasionally ”target sign” with a hyperechoic central region and a hypoechoic periphery, similar as MRI.
Pelvic myxoma	• Well-circumscribed • Low T1w and high T2w signal intensity • Variable heterogeneous enhancement with cystic nonenhancing areas	• Low-attenuating mass with cystic changes. • Mild enhancement of noncystic areas.	• Hypoechoic mass with cystic spaces and internal septae [20]
Aggressive angiomyxoma	• Hyperintense on T2w images • Isointense on T1w images • Strong enhancement with swirled central strands of hypointense tissue.	• Swirling internal architecture. • Contrast enhancing of fibrovascular strands similar to contrast enhanced MRI [8]	• Hypoechoic heterogeneous mass with cystic and solid components

Table 2: Differential diagnosis table of myxoid containing tumors with similar imaging features to aggressive angiomyxoma

ABBREVIATIONS

AAM = Aggressive angiomyxoma
MRI = Magnetic resonance imaging
R2 = indicates that portions of tumor visible to the naked eye were not removed.

KEYWORDS

aggressive angiomyxoma; abdominal imaging; tumor; MRI; histology; therapy; imaging

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