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Aggressive angiomyxoma of the pelvis: a series of four cases and literature review

Honglin Wu[1], Wei Liu[2], Hai Xu[3], Dehang Wang[3], Aimei Ouyang[4]

[1] Department of Radiology (H.L.W.), Affiliated Wujin Hospital of Jiangsu University, Changzhou, Jiangsu
[2] Department of Radiology (W.L.), Affiliated Changzhou Second Hospital of Nanjing Medical University, Changzhou, Jiangsu
[3] Department of Radiology (H.X.,D.H.W.), First Affiliated Hospital of Nanjing Medical University, Nanjing, Jiangsu
[4] Department of Radiology (A.M.O.Y.), Qilu Hospital of Shandong University, Jinan, Shandong (China)

Summary

The purpose of this study was to evaluate the value of CT and MRI in aggressive angiomyxoma (AAM) of the pelvis. A series of four cases from three institutions are reviewed. Among the four cases, three were initially misdiagnosed, and local recurrence necessitated reoperation or angiographic embolization. The fourth case, with accurate preoperative diagnosis, was followed with no recurrence. CT and MR imaging demonstrated a well-defined mass, which displaced adjacent structures. Attenuation of the mass was less than that of muscle on unenhanced CT, and a swirling or layering internal architecture was found using both enhanced CT and T1-weighted MR imaging. In one patient, a layering internal architecture was seen on unenhanced CT images. MRI demonstrated the relation of the tumor to the pelvic floor better than CT. The authors concluded that both CT and MRI show the characteristic imaging pattern and trans-diaphragmatic extent of these tumors, and the diagnosis should be considered in any young woman presenting with a well-defined mass arising from the pelvis or perineum.

Key words: Aggressive angiomyxoma; Pelvic organs; CT; MRI.

Introduction

Aggressive angiomyxoma (AAM) is an uncommon benign mesenchymal tumor preferentially arising from the connective tissue of the pelvic and perineum of adults. The tumor was first described in 1983 [1]. It has been found to affect women in more than 90% of cases, usually in the second to fourth decade. Pre-operative diagnosis is difficult, because it is often mistaken clinically for other benign conditions, such as Bartholin's cyst, leiomyoma, fibroepithelial polyp, lipoma, abscess, or hernia [2, 3]. Because of the high rate of local recurrence, preoperative CT or MRI is important to determine the extent and, thus, the optimal surgical management and complete excision. The authors present four cases, which illustrate the imaging features of AAM.

Case Report

Case 1

A 34-year-old woman presented with an eight-year history of increasing intermittent abdominal pain after a cesarean section. Ultrasonography revealed an asymmetrical mass. At surgery, a 12×3×3 cm mass was identified to the left of the rectum and the rectovaginal septum, closely adherent to the rectum and vagina. The tumor was completely removed and reported as AAM.

Three years later, the patient presented with a recurrent tumor. CT showed an 11×5×7 cm cystic mass with a characteristic swirled internal architecture in the left pelvis. During surgery, a soft mass was identified to the left of the rectum and the rectovaginal septum, adherent to the urinary bladder, vagina, and visceral pelvic fascia. The mass was removed as much as possible. The woman has since been on regular follow-up for five years with no recurrence of the disease.

Case 2

A 39-year old woman presented with a pelvic mass revealed by pelvic MRI during a routine medical examination. The lesion was found lying in the left ischio-rectal space (Figure 1A, B), although the patient had reported no symptoms. Then the patient underwent excision of the lesion via the transabdominal approach. During surgery, the tumor had a smooth surface and was found to be elastic and soft. The resected tumor was 15×8×3.5 cm, with histological examination confirming AAM.

After three months, a follow-up MRI demonstrated a 2.5×6 cm recurrent tumor in the same space as the original one (Figure 1C). Since the patient was unwilling to have another operation, she received an angiographic embolization. Subsequent imaging confirmed a significantly reduced tumor (Figure 1D). As the patient was asymptomatic, conservative management was undertaken.

Case 3

A 31-year-old woman presented with a vaginal wall cyst and had a vaginal wall cystectomy in another hospital in 1999. Four years later, she presented to the present institute with a recurrence. Physical examination revealed a 5×4 cm cystic mass in the anterior vaginal wall under abdominal pressure. The patient received an anterior vaginal tumor excision, and an AAM diagnosis was made.

After another six years, the woman presented to the present hospital again with a complaint of discomfort and swelling in the

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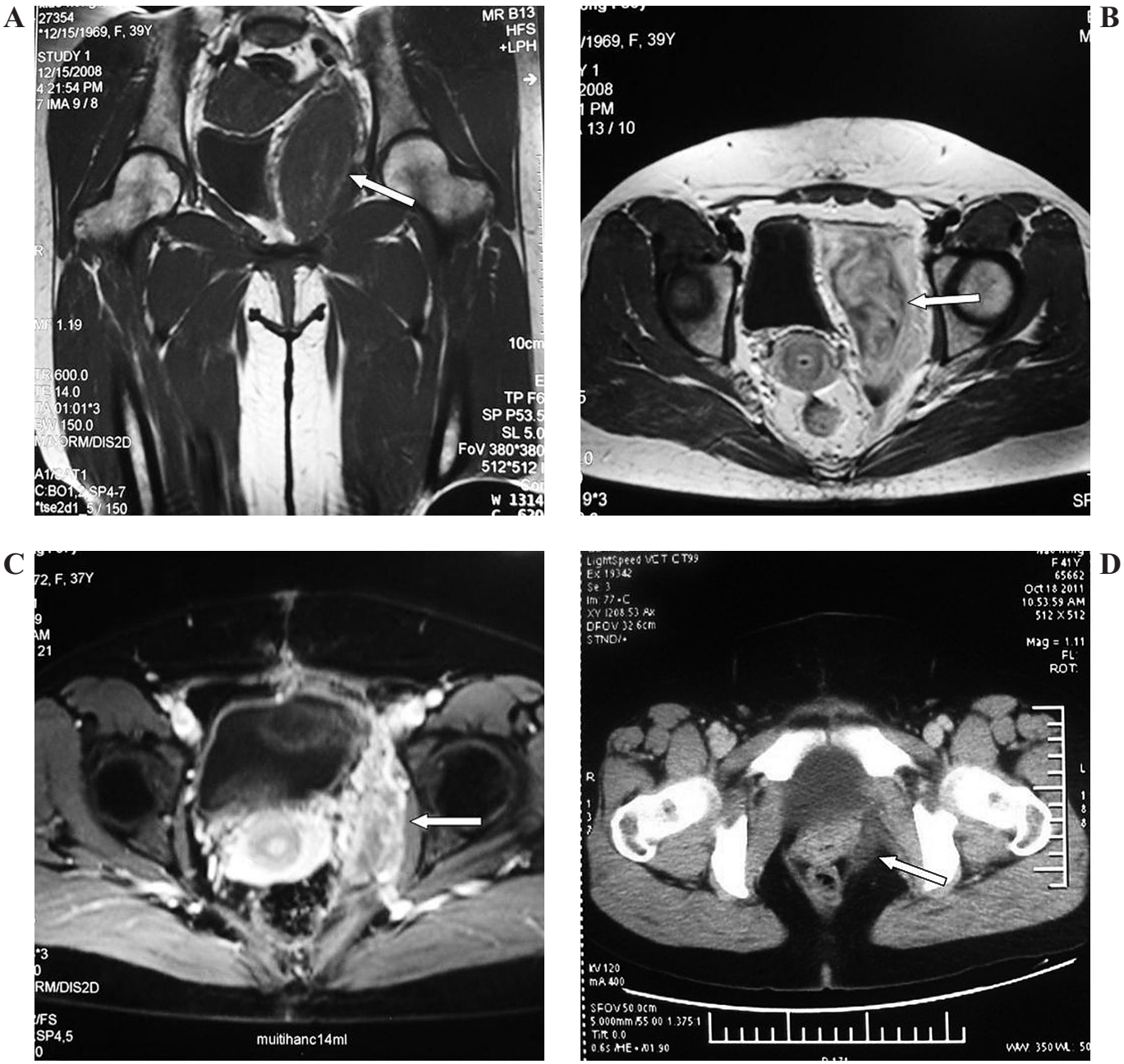

Figure 1. — A) Coronal unenhanced T1-weighted MRI shows an intrapelvic tumor (arrow), which is isointense to muscle and displaces the levator ani muscle, urethra, vagina, and rectum. B) Axial enhanced T1-weighted MRI showing marked tumor enhancement with swirled internal architecture. C) Axial enhanced T1-weighted MRI shows a recurrent enhanced tumor three months after surgery. D) Thirty months after angiographic embolization, CT shows a significant decrease of the tumor.

lower abdominal. On examination, a soft cyst measuring 10×10 cm was palpated in front of the uterus. Ultrasonography revealed an asymmetrical mass. CT showed a 10×16×17 cm mass traversing the pelvic diaphragm.

The patient underwent abdomino-perineal excision of the vulval and retroperitoneal mass. With an attempt to enucleate the mass, it was found that part of the mass was in the vagina with no obvious gap between the mass and the bladder. Subsequently, a portion of the bladder wall and vagina were resected, and as much of the mass as possible was removed. Histopathology was reported as AAM. Unfortunately, the patient was lost to follow-up because she never returned for a check-up.

Case 4

A 51-year old woman presented with a pelvic mass, detected during a routine medical examination. Color ultrasonography revealed a hypoechoic mass in the right pelvis. CT showed an intrapelvic mass above the pelvic diaphragm. Unenhanced CT

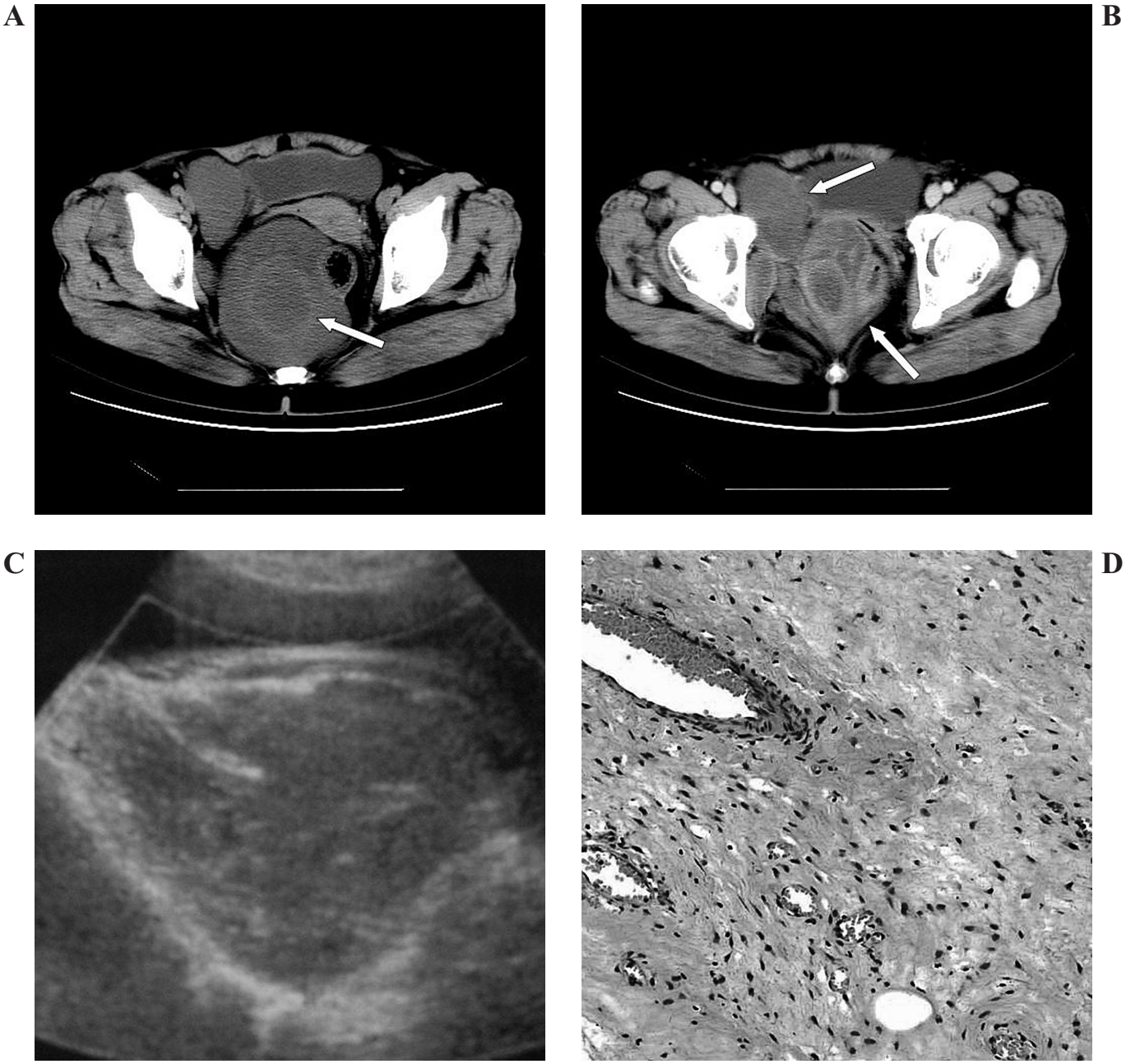

Figure 2. — A) Unenhanced CT shows layering internal pattern (arrow). B) Enhanced CT at a more caudal level shows that the tumor is polypoid and has an appearance of swirling strands, displacing the rectum, vagina, bladder, and levator ani muscle (arrow). C) Sonography reveals a hypoechoeic soft tissue mass that appears cystic. D) The tumor is characterized by hypocellular myxoid stroma with numerous variably sized blood vessels.

showed a hypo-attenuating mass with a layered internal architecture (Figure 2). Intravenous contrast material enhanced the strands within the tumor. According to the distinctive imaging appearances, a diagnosis of AAM was suspected preoperatively.

At surgery, the mass was located to the right of the rectum and the rectovaginal septum, closely adherent to the rectum and pelvic floor. Wide excision of the tumor was done and reported as AAM. The patient has been followed for eight months and is free from recurrence of the disease.

Discussion

AAM of the perineum or pelvis is a rare condition which predominately occurs in women of reproductive age, exhibiting a peak incidence in the fourth decade of life [4]. AAM typically arises mainly from the connective tissues of the perineum or lower pelvis, rather than directly from the pelvic viscera [5]. Owing to its slow

growth pattern, patients are often asymptomatic, and the tumors are often discovered incidentally during routine pelvic check-up or with radiographic imaging [4]. Although it is histopathologically categorized into benign neoplasm, the tumor is still called aggressive because of its high rate of local recurrence, where it can even invade the bladder, bowel or pelvic bone. AAM is generally a locally aggressive tumor, as distant metastasis has only been reported in three cases [2, 6].

Histopathology is definitely the gold standard for diagnosis of AAM. On gross examination, AAM is unencapsulated. It is soft to rubbery, poorly circumscribed, and may have finger-like projections that extend into neighboring tissues [7]. The histological appearance of AAM is characterized by a mixture of stellate or spindle cells within a strong myxoid background of collagen fibers and small vessels with thick walls [8]. Mitotic figures are unusual but are not atypical if present [2]. No specific immuno-histochemical marker has been found for AAM. Although most AAMs express estrogen receptor, progesterone receptor, desmin, vimentin, and smooth muscle actin, and is invariably negative for S-100, they are usually of little assistance in distinguishing AAM from its diagnostic mimics [9].

Ultrasound imaging usually reveals a hypoechoic soft tissue mass that may even appear cystic [10]. On sonography, it is homogeneously hypoechoic and well demarcated with multiple thin, echogenic internal septa [11]. There are several distinctive radiological features of AAM on CT and MRI that have been identified in the literature [12]. On cross-sectional imaging, AAM typically appears as a well-defined mass, displacing rather than invading adjacent organs with preservation of fat planes as shown in the present cases. The tumor usually traverses the pelvic diaphragm. CT appearances are variable, including a well-defined homogenous mass which is hypodense relative to muscle, a hypo-attenuating solid mass with a contrasting swirling internal pattern or a predominantly cystic appearing mass with a solid component. In case 4 of the present series, the internal layering pattern was seen clearly in enhanced CT. MRI is the best radiological modality for diagnosis. Characteristic appearances on MRI include iso-intensity relative to muscle on T1-weighted images and hyper-intensity on T2-weighted images, with avid and heterogenous contrast enhancement after administration of intravenous contrast, highlighting a distinct swirling or layering internal pattern. The hypo-density on CT and the hyper-intensity on T2-weighted MRI are likely due to the tumor's abundant loose myxoid matrix and high water content, while the avid enhancement reflects its high vascularity. The appearance of swirling or layering strands in the tumor is a characteristic diagnostic feature which is found in about 83% of the patients [13]. With CT, the swirled appearance is usually evident only with enhanced scans, but a layering internal architecture was seen on unenhanced CT images in the fourth patient. These are of slightly lower intensity than the remainder of the tumors in T2-weighted-MRIs and are also obvious after enhancement with gadolinium. The cause of this appearance is not very clear but may relate to the fibrovascular stroma that develop in tumors that are stretched as they protrude through the pelvic space or diaphragm (Figure 2). Aside from accurately diagnosing the tumor, MRI can be more helpful than CT in planning the surgical approach by defining the relation of the tumor with the anal sphincter, urethra, bladder and pelvic side-wall, and in clinical follow-up of patients with a recurrent tumor.

The first line of therapy for AAM is complete surgical excision with tumor-free margin, although achieving resection of tumor-free margins is difficult, owing to the infiltrative feature of the tumor and the absence of a definite capsule [2]. However, because of the rarity of AAM, there are no national or specialty guidelines for its management [14]. The high recurrence rate may be due to incomplete surgical excision, which may in turn be attributed to an incorrect initial diagnosis. In the present report, the first three patients were misdiagnosed and as a consequence, had recurrence within three months to six years. The fourth patient, in whom the preoperative diagnosis was correct, underwent a complete excision of the tumor with no recurrence. A review of more than 100 cases, however, refuted this by showing that patients with clear resection margins were just as prone to developing recurrences [15], and an incomplete removal is supported by recent literature in vulvar AAM [16].

Other treatment modalities for AAM include chemotherapy, radiation therapy, angiographic embolization, chemoembolization, and hormonal therapy. These are sometimes used as adjuvant therapies for a residual tumor, in the treatment of recurrence, or as a help in succeeding resection by shrinking the neoplasm, and making it easier to distinguish from surrounding normal tissues [3,17]. In case 2, subsequent imaging showed that the tumor gradually shrunk significantly. However, chemotherapy and radiotherapy are usually thought to be less suitable options because of their low mitotic activity, and angiographic embolization is also controversial [18]. Furthermore, an alternative blood supply may cause a recurrence after the initial response to embolization, necessitating a longer follow-up observation period. AAMs in women are reportedly hormone-responsive tumors. Hormonal therapy is found to have a significant role in the treatment of extensive or recurrent AAM that is estrogen receptor positive. However, hormonal therapy also have long-term adverse effects including menopausal symptoms and bone loss [14].

In summary, AAM is a rare tumor that affects the pelvis or perineum. The tumor has a distinctive imaging appearance of swirled or layered internal architectures after enhancement on CT and MRI. Surgery is potentially curative with recurrence usually due to incomplete resection. The multiplanar images of CT and MRI can reveal extension of

the tumor and are thus valuable in determining the optimal surgical approach.

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Address reprint requests to:
HONGLIN WU, M.D.
Department of Radiology
Affiliated Wujin Hospital of Jiangsu University
Yongning No. 2 North Road, Tianning District
Changzhou, Jiangsu 213002 (China)
e-mail: wuhljs@163.com