MR Imaging of Aggressive Angiomyxoma of the Female Pelvis: Case Report

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Aggressive angiomyxoma is a rare tumor that predominantly occurs in the female genital tract. Because of its high tendency for local recurrence, preoperative diagnosis is important to ensure wide excision and reduce recurrence risk. But it is often misdiagnosed due to its rarity. We present two cases of aggressive angiomyxoma: a recurrent case that was initially misdiagnosed and a preoperatively diagnosed case.

Index words : Myxoma
Magnetic resonance (MR)
Vulva
Female
Perineum

Introduction

Aggressive angiomyxoma is a recently characterized benign mesenchymal tumor occurring predominantly in the perineal area, with a strong female predominance in the 3rd-5th decades of life (1). It has a high propensity for local recurrence, thus preoperative diagnosis is important for planning surgical management (2). But it has been often misdiagnosed due to its rarity (3).

To our knowledge, the imagings of aggressive angiomyxoma have not been reported in Korea. We present here our experience of a local recurrence case which was initially misdiagnosed and another case which was correctly diagnosed preoperatively.

Case 1

A 43-year-old woman presented with a non tender mass in the right perirectal area in 1999. Magnetic resonance (MR) imaging showed a large soft-tissue, 12 $\times 8 \times 5$ cm mass, mainly located at the right perineum extending to the perirectal space. The mass displaced the adjacent soft tissue and splitted the internal and external right levator ani sphincter muscle. It was isointense to muscle on T1-wighted and hyperintense on T2-weighted images, with a whirling pattern (Fig. 1). Differential diagnosis included a neurogenic tumor, myxoid liposarcoma, angiomyolipoma or other soft tissue origin tumors.

Local excision was performed. Gross examination demonstrated a soft and gelatinous mass. Microscopically, it was composed of smooth muscle

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cells in myxoid background with prominent thick-walled vessels; mitotic activity was absent. Histologic diagnosis was angiomyolipoma or myxoid smooth muscle tumor.

One year later, a small recurrence was noted in the right perineum on CT scan which slowly continued to grow for 8 years, until the patient finally presented with increased swelling and pain of the right perineum. On MR imaging, the imaging findings were similar to the initial presentation. Internal texture showed high signal intensity interspersed with swirled strands of lower intensity on T2-weighted images and showed heterogenous enhancement (Fig. 1). At this time radiologic diagnosis of an aggressive angiomyxoma was made. Complete excision was performed, which revealed a lobulated $10 \times 8 \times 5$ cm tumor in the right ischiorectal fossa. Pathological diagnosis was aggressive angiomyxoma (Fig. 1).

Case 2

A 44-year old woman presented with a perirectal mass. MR imaging showed a solid and cystic mass that extended from the left ischiorectal fossa to the pelvis, displacing the levator ani muscles anterosuperiorly, and

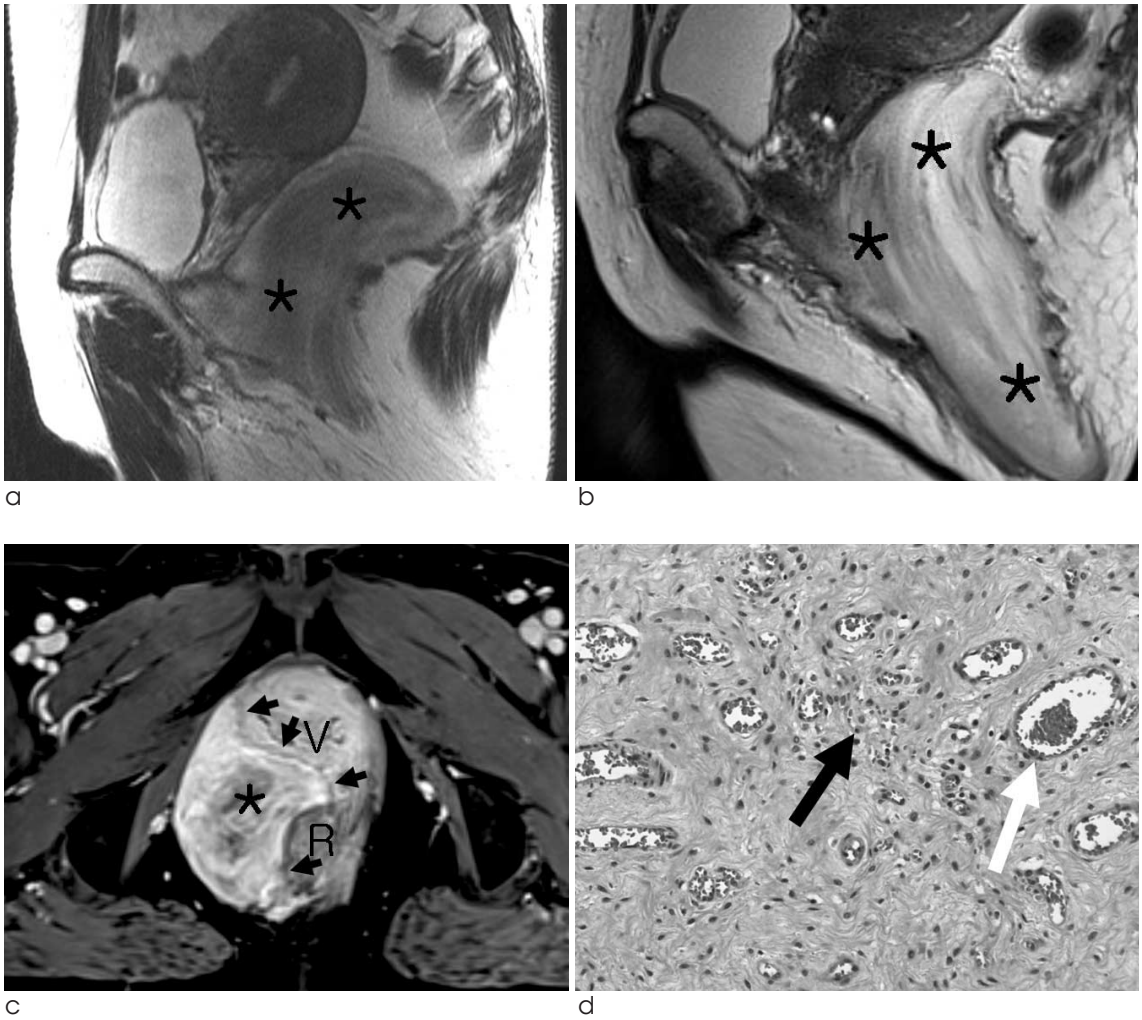
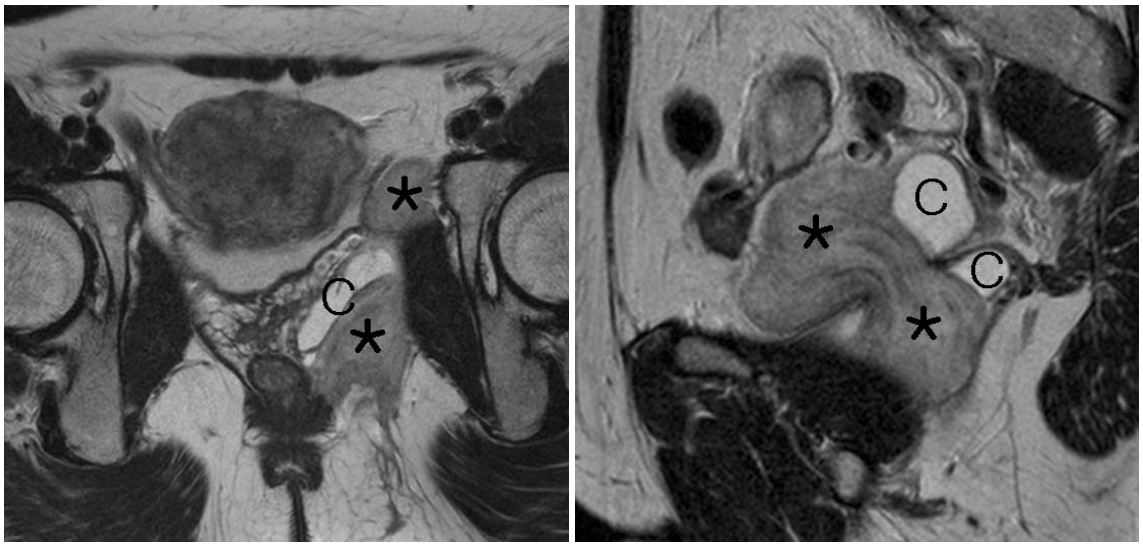


Fig. 1. 43-year-old female with aggressive angiomyxoma extending from perineum to pelvis.
a. T2-weighted fast spin-echo (FSE) imaging (TR/TE = 4,000/98). Sagittal image shows right perineal mass (asterisks) extending through pelvic diaphragm displacing the uterus and a swirling appearance within the tumor. b. Recurred tumor 8 years after excision. Sagittal T2-weighted FSE imaging (TR/TE = 5,310/113) show a similar appearance to that of the primary lesion (asterisks). c. Axial gadolinium-enhanced T1-weighted spin-echo with fat suppression imaging shows a swirling pattern of heterogeneous enhancement within the tumor (asterisks and black arrow). V = vagina, R = rectum. d. Histological section shows spindle and stellate cells in a myxoid background with scattered dilated bloods vessels in small and middle sizes (white arrow) and extravasated red blood cell (black arrow). Hematoxylin-eosin stain: $\times 100$.



a b

Fig. 2. A 44-year-old female with aggressive angiomyxoma extending from vulva to pelvis.
a, b. T2-weighted FSE imaging (TR/TE = 2,887/80). a. Oblique axial image (asterisks) shows a solid and cystic mass arising from the left ischiorectal fossa displacing the rectum. b. Sagittal image shows the tumor with swirling strands. C = cystic areas within the mass.

the rectum and vagina towards the right. On T2-weighted images the tumor had a swirling internal architecture (Fig. 2). Based on the imaging findings, radiological diagnosis of aggressive angiomyxoma findings was suggested. Wide excision was performed and aggressive angiomyxoma was confirmed. The patient remains disease-free at 24 months.

MR imaging sequence

MR examinations were performed on a 1.5-T (Genesis sigma, General electric Medical Systems, Milwaukee, WI. USA or Intra achieva, Philips Medical systems, Eindhoven, Netherlands) or 3-T imaging system (Triotim, Siemens, Erlangen, Germany). The patients underwent the following sequence imaging axial, saggital, oblique axial and oblique coronal T2-weighted fast spin echo (FSE) sequence (TR/TE = 2,887–5,000/80–113 msec; echo-train length, 12-35, flip angle, 90°–140°; matrix, 320 × 320; section thickness, 4–5 mm; intersection gap, 4–6.5 mm), and axial T1-weighted spin echo sequence (TR/TE = 539/12 msec; flip angle, 90°; matrix, 320 × 320; section thickness, 5-6 mm; intersection gap, 7–9 mm). Precontrast and Gd-BOPTA (Dotarem, Guerbet, Rossy, France)-enhanced images were obtained using the T1-weighted volumetric interpolated breath hold examination (VIBE; TR/TE = 11.2/4.39 msec; flip angle, 10°; matrix, 320 × 320; section thickness, 1 mm). 0.1 mmol Gd-BOPTA per kilogram of body weight was given to the patient intravenously.

Discussion

Aggressive angiomyxoma is a rare tumor found predominantly in the female genital tract (1). It grows slowly and is often large at the time of diagnosis (usually larger than 10 cm) (2). Surgical resection is the main treatment modality. However, because of infiltrative behavior, the local recurrence rate is 36 to 72% and 70% recurrence within a two year period (2). As a result, the widest excision technically possible is warranted. To ensure this, accurate preoperative diagnosis via imaging is important.

The characteristic MR imaging appearance of angiomyxoma may aid in the differential diagnosis. MR imaging features have been previously described as iso- or hypointensity on T1-weighted and hyperintensity on T2-weighted images (4). The high signal intensity on T2-weighted images reflects the high water content and myxoid matrix. A whorled pattern of signal intensity on T2-weighted images has been reported as a characteristic feature (4). These tumors also show contrast enhancement that reflects inherent vascularity. Infiltrative patterns are not usually seen, and tumors

tend to displace and grow around adjacent structures. These findings are consistent with the findings in our cases.

The differential diagnosis of a pelvic or perineal soft tissue mass in an adult female paitent includes angiomyofibroblastoma, myxoma, infiltrating angiolipoma, and myxoid lipoma. Angiomyofibroblastoma, a type of angiomyxomas, may be confused with aggressive angiomyxoma. Angiomyofibroblastoma is usually small and affects the superficial vulva and vagina, whereas an aggressive angiomyxoma involves deep tissue planes and is often large at the time of diagnosis (5). Myxoma, another benign mesenchymal neoplasm, is mainly located intramuscularly in the thigh and pelvic girdle, and frequently occurs in older patients. Aggressive angiomyxoma may abut the pelvic or perineal musculature, but does not invade it. Microscopically, myxoma has negligible vascular components and does not enhance, as does aggressive angiomyxoma (6). Infiltrating angiolipoma is also a rare benign soft tissue tumor similar to aggressive angiomyxoma in that it has tendency for local recurrence requiring wide surgical excision. However, infiltrating angiolipoma usually found in the extremities and extensively infiltrates muscle, fascia, and subcutaneous tissue. It has a characteristic appearance on computed tomography (CT), which is a largely intramuscular tumor with a heterogeneous appearance reflecting fat and blood vessels within infiltrated muscle (7).

Myxoid lipoma is composed of myxoid and fat, myxoid component predominating. But it is different from aggressive angiomyxoma in having mature fat with a trasitional zone between the myxoid and fat components (1). Myxoid liposarcoma commonly occurs in lower extremities. Myxoid liposarcoma demonstrate a spectrum of MR imaging. Most myxoid liposarcomas demonstrate lacy or linear, amorphous foci of fat. It shows homogenous enhancement with contrast, whereas aggressive angiomyxoma is more heterogeneously enhanced (8).

In the first case, we did not have knowledge about aggressive angiomyxomas at the initial time of MR imaging interpretation. We were unable to correctly diagnosis the tumor, and it seemed to be incompletely excised. The local recurrent mass grew very slowly over 8 years, had similar appearance to the primary lesion. It remained well defined, and displaced adjacent structures. It is consistent with previous report (9). In the second case, we were able to correctly diagnose aggressive angiomyxoma with imaging (4, 10). The patient underwent wide excision and remains disease free at 24 months.

In conclusion, aggressive angiomyxoma can be misdiagnosed because of its rarity and radiologists' lack of familiarity with its imaging findings. However, it has characteristic appearances on MR imaging in both primary and recurrent cases. Knowledge of these features may help to achieve correct diagnosis of vulval and perineal masses, and lead to proper treatment.

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여성 골반에 생긴 공격성 혈관점액종의 자기공명영상: 증례 보고

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공격성 혈관점액종은 여성 생식길에 주로 생기는 드문 종양으로 국소 재발율이 높다. 광범위 국소 절제술을 통해 재발 위험을 줄일 수 있기 때문에 수술 전 진단이 중요하다. 그러나, 이 종양이 드물어 오진되는 경우가 종종 있다. 우리는 수술 전 오진으로 재발한 공격성 혈관점액종 증례와 바르게 진단한 두 증례를 경험하여 보고한다.

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