Aggressive Angiomyxoma Extending Largely into the Pelvis
—— A Case Report and Review of the Literature ——

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Aggressive angiomyxoma (AA) is a rare soft tissue tumor that involves the vulvar and perineal regions of afflicted individuals. It is a slow-growing and locally infiltrating tumor that is characterized by an increased risk of local recurrence. In this report, we describe the case of a large AA in a 39-year-old woman. She was admitted to our hospital with a pelvic tumor, which was discovered during a gynecologic checkup, and magnetic resonance imaging revealed its presence traversing from the pelvic diaphragm to the perineum. The tumor was completely excised using an abdominal approach, and the pathological diagnosis was that of AA. The patient had an uneventful recovery and no evidence of recurrence exists 12 months after surgery. Long-term follow-up and careful monitoring, however, are necessary because AA is slow-growing. As AA is usually reported as a vulvar tumor, the present case is a rare incidence in which the AA grew largely into the pelvic cavity. (Kitakanto Med J 2009 59: 157~160)

Key Words: Aggressive angiomyxoma, Pelvic tumor, Diagnosis, Treatment, GnRH agonist

Introduction
Aggressive angiomyxoma (AA) is a rare and distinctive soft-tissue neoplasm that chiefly involves the vulvar and perineal region of female patients. These lesions present as soft, non-encapsulated tumors with fingerlike projections that infiltrate the surrounding soft tissues displacing adjacent organs but not typically invading the adjacent organs. AA was first described by Steeper and Rosai1 in 1983, and approximately 200 cases have been reported in the English literature. Most cases of AA are reported as vulvar tumors. Here, we report a case of AA that largely extended into the pelvis and did not present in the form of a vulvar tumor.

Case Report
A 39-year-old asymptomatic woman was admitted to our hospital with a pelvic tumor that was detected during a gynecologic checkup. No evidence was seen of tumor formation in the vulvar or perineal region. She was healthy, with no remarkable past medical or family history. Biochemical and peripheral blood tests were normal. Tumor marker levels for carcinoembryonic antigen, carbohydrate antigen 19-9, and cancer antigen 125 were within their normal ranges. A transvaginal ultrasound scan revealed a giant homogeneous and hypoechoic mass with poorly defined margins. A computed tomography (CT) scan of the pelvis detected a large hypoattenuating tumor, which was indistinguishable from the left rectal wall (Figure 1A). The lesion was displacing the rectum, bladder, and uterus toward the right side of the pelvis (Figure 1B). Magnetic resonance imaging (MRI) showed a large tumor in the pelvis with the tumor traversing the pelvic diaphragm to the perineum, exhibiting a characteristic “swirled” internal pattern measuring 20×12 cm (Figure 2).

Following diagnosis of the pelvic tumor, the patient underwent surgery and had the tumor removed.

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Figure 1  CT of the pelvis.
A: The tumor was indistinguishable from the left rectal wall.
B: The lesion was displacing the rectum, bladder and uterus toward the right side of the pelvis.

Figure 2  MRI showed a large tumor in the pelvis with the tumor traversing the pelvic diaphragm to the perineum with a characteristic “swirled” internal pattern, measuring 20×12 cm.

Figure 3  A: The tumor was 30×10×6 cm in size and weighed 740 g.
B: The cut surface was glossy, gray-white and solid.

Figure 4  A: The tumor was composed of spindle and stellate cells and blood vessels with thick walls distributed in an abundant myxoid stroma (H-E ×100).
B: The tumor cells exhibited positivity with estrogen receptors (×100).
The tumor was soft and rubbery, and was indeed found to be displacing the rectum, bladder, and uterus toward the right side of the pelvis. Although no invasion of the adjacent structures was present, the border of the tumor and the surrounding tissue was indistinct. Additionally, the tumor was indistinguishable from the rectal wall. We completely resected the tumor by using only an abdominal approach. The tumor was $30 \times 10 \times 6$ cm in size and weighed 740 g (Figure 3A). The cut surface exhibited a glossy, gray-white appearance and was solid in nature (Figure 3B). Histological examination of the tumor revealed AA. The tumor was composed primarily of spindle and stellate cells and blood vessels with thick walls distributed in an abundant myxoid stroma (Figure 4A). Immunohistochemical analysis determined that the tumor cells were positive for estrogen and progesterone receptors (Figure 4B). No nuclear pleomorphism or mitotic activity was detected.

The postoperative course was uneventful. No evidence of tumor recurrence has emerged 12 months following surgery.

**Discussion**

AA was first reported as a distinct variant of myxoid neoplasms in the female pelvis and perineum by Steeper and Rosai. It is a characteristically infiltrative, locally aggressive, nonmetastatic fibromyxoid soft tissue tumor. The reported female-to-male ratio is 6.6 : 1. AA occurs across a wide age range from 7 to 70 years, with peak incidence occurring during the reproductive years. In women, the majority of AA occurs in the vulva and perineum. Although some cases have been reported in which tumor growth progresses into the pelvis from the vulva, the present case is rare since AA did not arise in the form of a vulgar tumor and yet tumor growth progressed largely into the pelvis. In a search of the English language literature from 1983 to 2007, we only found about 20 similar cases in which a large tumor of this type was noted. AA is a complex tumor that is locally aggressive and infiltrative, but tends to only displace adjacent organs without invading them. Because of the slow, insidious growth pattern of these tumors, patients are often asymptomatic and visible vulvar masses or tumors are discovered incidentally during pelvic examinations or imaging studies. It is often misdiagnosed as a Bartholin's duct cyst, lipoma, or vaginal cyst. Histological examination shows a predominant myxoid stroma and an abundance of thin and thick-walled vascular channels, and cellular mitosis is absent in most cases. Pathological differential diagnosis is typically angiomyofoibroblastoma, superficial angiomyxoma, and smooth muscle tumor with remarkable myxoid change. The pathogenesis is unclear, but a translocation at chromosome 12 with a subsequent aberrant expression of high-mobility-group protein-2 (HMGA2), which is involved in DNA transcription, has been recently demonstrated. Imaging patterns are atypical since each case of AA varies depending on the level of differentiation present in the tumor cells. AA has been reported to display iso- or hypooattenuation relative to muscle on CT. Moreover, it is isointense relative to muscle on T1-weighted images, hyperintense on T2-weighted images, and enhanced greatly after gadolinium contrast, displaying a characteristic “swirled” internal pattern on MRI. High signal intensity on T2-weighted MR images may be caused by the myxomatous stroma of these tumors.

The usual course of treatment is complete excision of the tumor. However, because of the locally aggressive and infiltrative nature of this type of tumor growth, recurrence rates as high as 36 to 72% have been reported even when the tumor exhibits clear surgical margins. More recent studies have reported lower recurrence rates of only 9%, but these have had limited follow-up periods of only 12 to 16 months. Since this is a slow-growing tumor and recurrence has been reported 14 years after initial tumor removal, long-term follow-up care is necessary. High recurrence rates are common in cases when no distinctive margins occur, which results in an indistinguishable border from the adjacent organs. In cases of an incomplete resection, an extended resection may be needed to prevent the loss of adjacent organs. Currently, incomplete resection is deemed acceptable only when preservation of fertility is desired and high operative morbidity from an extensive surgical procedure is anticipated.

Because AA exhibits low mitotic activity, radiation or chemotherapy are not seen as appropriate treatment options. Several studies have demonstrated that the cells found within an AA mass possess estrogen and/or progesterone receptors, suggesting that AA is a hormonally responsive neoplasm. Cases of recurrent and residual tumors have been successfully treated with gonadotropin-releasing hormone (GnRH) agonist. In addition, this hormonal treatment has the possibility of being used as a neoadjuvant therapy to reduce tumor size and preserve organs, and as adjuvant therapy after incomplete resection. However, one study has reported a recurrence after completion of effective adjuvant hormone therapy. Problems arise when hormone therapy is continued for extended periods of time because GnRH agonist treatment might induce several side effects. One proposed idea is to pursue an intermittent administration of GnRH agonist. Additionally, several studies have
reported a relatively low recurrence rate in patients over 50 years of age; thus, the possibility exists that the progression of AA may be halted at the onset of menopause, allowing the discontinuance of hormonal therapy.

Even though one of the distinguishing characteristics of AA is a high local recurrence rate, AA is generally regarded as a benign tumor. However, two cases with distant metastases and associated tumor-related deaths have been reported. Therefore, AA must be recognized as having a metastasizing potential.

Here, we report a large AA that was successfully removed following surgical resection. No evidence of recurrence is present 12 months after surgery, but because AA is slow-growing, long-term follow-up and careful monitoring with imaging techniques are necessary.

References


