Huge Aggressive Angiomyxoma: A Case Report and Literature Review

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Angiomyxoma occurs mostly in 30- to 40-year-old females and is described histologically as a mesenchymal tumor, composed of fibroblasts within a strong myxoid background. It occurs mainly in the female pelvis, vulva or perineum, and grows slowly. Treatment is surgical excision. Unfortunately, there is a relatively high rate of recurrence because the exact extent of the tumor is difficult for the surgeon to determine. We report a case of aggressive angiomyxoma combined with uterine myoma, and discuss the characteristics of its images. Surgical excision of the tumor was performed, and adjuvant treatment was given for local recurrence.

Key Words: aggressive angiomyxoma, female, diagnosis, treatment

In 1983, Steeper and Rosai described aggressive angiomyxoma, primarily in the female pelvis and perineum, as a distinctive variant of myxoid tumors with prominent vascular component [1]. The tumor occurs predominantly in the premenopausal female. We present a case of aggressive angiomyxoma of the pelvis that coexisted with uterine myoma. After surgical resection, the patient was followed-up in our outpatient department; if local recurrence occurs, she will be treated with gonadotropin releasing hormone (GnRH) agonist. A review of the literature is also outlined.

CASE REPORT

A 39-year-old unmarried, nulliparous woman presented with a history of nonspecific abdominal symptoms and a right lower abdominal mass for 2 weeks. She came to our gynecologic clinic for help. Abdominal sonography showed a solid mass with heteroechochogenicity in the pelvic cavity. Abdominal computed tomography (CT) showed a huge cystic mass in the pelvis and uterine myoma (Figure 1). Magnetic resonance imaging (MRI) also revealed a huge soft tissue mass (18.5 × 7.8 × 11 cm) in the pelvic cavity, which showed low signal intensity on T1-weighted MRI, high signal on T2-weighted MRI with heterogeneous enhancement after contrast administration (Figure 2). Some foci showed strong enhancement within the mass. Uterine myoma was also found. The diagnosis of retroperitoneal liposarcoma or right ovary cystadenocarcinoma was favored by our radiologist and biopsy was recommended. After discussion with the patient, we decided to perform radical tumor excision due to high malignant potency.

During exploratory laparotomy, a huge teardrop-shaped mass was found in the pelvis, with invasion of the levator ani. Extended tumor excision and myomectomy were performed smoothly. Histologic examination of the specimen showed a myxoid tumor with sparse infiltrates of polymorphic cells clustering around blood vessels. Immunohistochemical studies showed strong staining for estrogen and progesterone receptors (Figure 3). Staining for desmin and vimentin was intermediate, and staining for S-100 protein was negative. The histologic diagnosis was aggressive angiomyxoma.
After the operation, no stool or urine incontinence was noted. The patient recovered well and returned to normal daily activity. Pelvic CT (Figure 4) was performed in our outpatient department after 6 months, and revealed no evidence of local recurrence. If local recurrence does occur, adjuvant hormone therapy with GnRH agonist will be given.

**DISCUSSION**

Aggressive angiomyxoma is a mesenchymal tumor that affects women in >90% of cases [2,3]. The tumor, which is composed of scattered spindle cells and abundant medium-sized vessels embedded in a myxoid matrix [1], arises from connective tissues of the perineum or lower pelvis, but rarely arises directly from any pelvic or perineal viscus [4,5]. The lesions are characterized by a predominantly myxoid pattern, different modes of presentation and locally infiltrative behavior with tendency for multiple recurrences.

Two factors may contribute to the high rate of local recurrence. First, the tumor is not diagnosed before initial surgery, and the extent of the tumor is frequently not perceived. Second, the strategic location of the tumor around...
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the urethra, vagina, and sphincter, and the rectum — with extension above and below the pelvic diaphragm — makes complete resection difficult and does not lend itself to either abdominal or perineal surgical approaches alone. For these reasons, most recurrences are likely to be related to inadequate resection and residual tumor [6].

Imaging studies of these tumors are important to determine the extent and optimal surgical approach. Outwater et al found that the characteristics of angiomyxoma has some variation on CT and MRI [7]. On CT, the tumor has a well-defined margin and attenuation less than that of muscle. On T2-weighted MRI, the tumor has high signal intensity. The attenuation on CT and high signal intensity on MRI are likely to be related to the loose myxoid matrix and high water content of angiomyxoma [7]. Similarly, we also found this tumor to have a swirled appearance on enhanced CT (Figure 1). CT and MRI can accurately reveal whether a tumor transverses the pelvic diaphragm. This determination is critical to choose a surgical approach: perineal or abdominal or both.

The tumor occurs predominantly in the premenopausal female, suggesting that estrogen may stimulate its growth. This concept is supported by a previous case report of rapid growth of an angiomyxoma during pregnancy [8], a state of increased estrogen and progesterone production. The success of adjuvant therapy with a GnRH agonist following complete surgical excision of an angiomyxoma [9] gave us another choice if local recurrence should occur. The patient’s aggressive angiomyxoma is clearly a dramatic example of an estrogen and progesterone receptor-positive tumor. Uterine myoma existing in this nulliparous female also indicates hormonal imbalance. Therefore, follow-up for local recurrence was mandatory for this patient, and adjuvant GnRH agonist will be given if local recurrence should occur.

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侵襲性血管黏液瘤 (aggressive angiomyxoma) 可發生於全身軟組織。臨床多見於 30—40 歲以上女性，可見於外陰、陰道黏膜下和盆腔軟組織，腫瘤可較大，生長慢，對骨骼肌和脂肪組織有一定的侵襲性。治療原則是完全切除腫瘤，但是局部復發常見。我們提出一個少見的侵襲性血管黏液瘤合併子宮肌瘤的案例，探討其影像學的診斷特徵，以手術的方式，成功的去除腫瘤，同時並討論如何以術後的藥物控制來治療及預防局部復發的產生。

關鍵詞：侵襲性血管黏液瘤，女性，診斷，治療

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