



Aggressive Angiomyxoma of the Abdominopelvic Cavity: A Case Report

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Abstract

Background: Aggressive Angiomyxoma (AAM) is a rare type of mesenchymal tumor that affects the pelvic and perineal regions of premenopausal women. AAM is a slow-growing, locally infiltrating tumor that tends to recur locally but is unlikely to metastasize. To date, surgery is the standard treatment for AAM, although it carries limited success rates. The literature around AAM is also limited.

Case Presentation: We report an unusual case of a 67-year-old woman with an extensive soft-tissue mass in the pelvic and abdominal cavities. We performed complete tumor excision, and a diagnosis of AAM was confirmed via pathology. The mass recurred within 5 months of surgery and grew in size rapidly. A wider tumor excision was performed 8 months after the initial surgery; however, a recurrent mass was again detected during the subsequent follow-up period. Four months after the second resection, the patient underwent 1 cycle of chemotherapy (cisplatin, ifosfamide, and epirubicin) at another hospital; however, the diameter of the mass continued to increase. At our last follow-up visit (15 months after the initial surgery), the patient was in poor health and had abandoned further treatment.

Conclusion: AAM should be considered in the differential diagnosis of asymptomatic, slow-growing masses in the abdominopelvic cavity. Wide local excision and long-term follow-up are essential for treating AAM.

Keywords: Aggressive angiomyxoma; Mesenchymal tumor; Wide local excision

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Received Date: 09 Jan 2017

Accepted Date: 06 Feb 2017

Published Date: 08 Feb 2017

Citation:

Yun-jie Tian, Kang S, Li L, Xi-wa Zhao. Aggressive Angiomyxoma of the Abdominopelvic Cavity: A Case Report. *Ann Clin Case Rep.* 2017; 2: 1263.

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Introduction

Aggressive angiomyxoma (AAM) was first described by Steeper and Rosai in 1983 as a distinct mesenchymal tumor of the pelvis and perineum in women [1]; it was described as “aggressive” to emphasize the neoplastic nature of the blood vessels and its locally infiltrative and recurrent nature.

Fewer than 250 cases of AAM have been reported in the literature, most of which occurred in women [2]. A recent literature review that assessed over 100 cases of AAM calculated a female-to-male incidence ratio of 6.6:1 [2]. Women who develop AAM are predominantly of childbearing age; the peak incidence is in the fourth decade of life, although the age distribution (ranging from 6 to 77 years) is wide [1-6]. AAM occurs predominantly in the vulvovaginal, perineal, and groin regions; those that occur in men involve analogous sites, including the inguinoscrotal region and perineum [7].

Surgical excision is generally recommended for the management of AAM. In 1992, Simo et al. [8] proposed that curative treatment for AAM should involve wide surgical excision based on precise histopathological diagnoses. Here, we report a rare case of a 67-year-old woman with AAM in the pelvic and abdominal cavities.

Case Presentation

A married 67-year-old woman (gravida 2; para 1) complained of progressive abdominal distension and anorexia. Upon physical examination, her abdomen was soft and distended. Gynecological examination revealed a 200 × 100 × 100 mm mass with low mobility and without clear margins. Ultrasonography revealed an irregularly shaped soft-tissue tumor with mixed echogenicity, approximately 195 × 129 × 108 mm in size (Figure 1). Computed Tomography (CT) of the abdomen and pelvis revealed a huge, hypodense mass in the pelvic and lower abdominal regions with a longitudinal diameter of 190 mm (Figure 2). The patient had a normal menstrual



Figure 1: Transabdominal ultrasonography revealed an irregularly shaped, soft-tissue tumor with mixed echogenicity (arrow).



Figure 2: Computed tomography (CT) of the abdomen and pelvis revealed a large, hypodense mass (arrows) in the pelvic and lower abdominal regions before the first surgery. (A) plain CT scan; (B) CT scan in the arterial phase; (C) CT scan in the venous phase.

history, no history of taking drugs, and had not undergone any prior surgical procedures. Surgical exploration revealed a soft, poorly circumscribed mass, approximately 200×150×100 mm in size, within the pelvis and abdomen. A spontaneous rupturing of the mass, which produced bloody ascites approximately 200 mL in volume, was also noted. The tumor had adhered to the intestines and the major omentum, and covered the uterus as well as the adnexa bilaterally. After careful dissection, the uterus and the adnexa appeared to be normal. The surfaces of the liver, spleen, stomach, and appendix were smooth. Owing to the high possibility of a mesenchymal tumor, we performed tumor resection, total abdominal hysterectomy, bilateral salpingo-oophorectomy, and omentectomy. On gross examination, there were large, poorly circumscribed lesions with irregular extensions into the surrounding tissues. Microscopic examination showed the proliferation of small fusiform or star-shaped cells without cytonuclear atypia that were interspersed in a myxoid background enclosing several capillary structures with thin walls (Figure 3). Immunohistochemistry showed that the tumor strongly expressed desmin and moderately expressed vimentin, CD99, CD34, and smooth muscle actin; the tumor was negative for CD117, cytokeratin, estrogen receptor, progesterone receptor, and S100. The diagnosis of AAM was confirmed on pathologic examination. After radical surgery, no further treatment was conducted except for close follow-up.

Five months after surgery, a recurrence of the mass was detected. Ultrasonography revealed a hypoechoic, solid tumor (71 × 56 × 42 mm) located on the vaginal stump. At the next monthly follow-up, magnetic resonance imaging (MRI) confirmed an irregularly shaped tumor in the pelvic cavity, measuring 84 mm in diameter (Figure 4). Eight months after the surgery, the recurrent mass was approximately

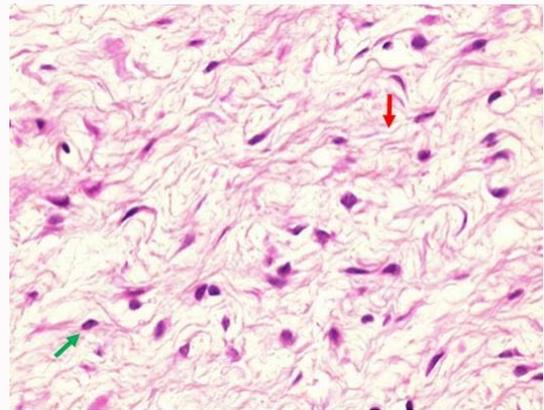


Figure 3: Photomicrograph (hematoxylin-eosin stain, original magnification ×40) showing the proliferation of small fusiform or star-shaped cells without cytonuclear atypia (green arrow) interspersed in a myxoid background (red arrow) enclosing several capillary structures with thin walls.

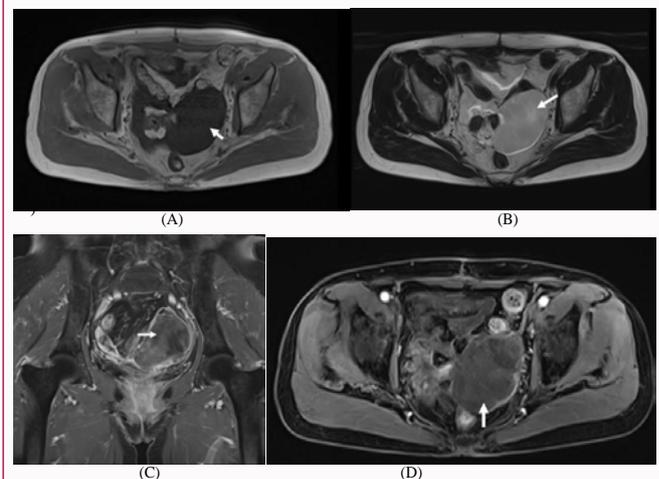


Figure 4: Magnetic resonance imaging (MRI) at first recurrence confirmed an irregularly shaped tumor in the pelvic cavity measuring 84 mm in diameter. (A) Axial enhanced T1-weighted MRI showed an aggressive angiomyxoma (arrow) that was isointense to the muscle; (B) Axial unenhanced T2-weighted MRI showed that the tumor was hyperintense relative to the muscle, and a swirling or layered appearance (arrow) was noted within the mass; (C) The tumor (arrow) was isointense on coronal enhanced T1-weighted imaging; (D) An axial enhanced T1-weighted image showed the tumor (arrow) with heterogeneous enhancement.

204 × 113 × 97 mm in size; the mass grew gradually to the point where it severely affected gastrointestinal peristalsis and caused abdominal distention. Hence, the patient underwent a second, wide excision. Surgical exploration found that the tumor in the pelvis and abdomen, 200 × 185 × 160 mm in size, was gelatinous, poorly circumscribed, and had no capsule. The recurrent tumor had occupied the inter-intestinal space and the vaginal stump. After the second operation, we continued close follow-up. Four months after the second tumor resection, a third recurrent mass developed. Ultrasonography revealed a solid, cyst-like mass in the pelvis and abdomen 294 × 272 × 100 mm in size. Following the third recurrence, the patient sought treatment at another hospital, where she was administered a chemotherapy regimen of cisplatin, ifosfamide, and epirubicin for 1 cycle. Five months after the second operation, ultrasonography indicated an irregularly shaped mass measuring 409 × 335 × 128 mm. When she was last seen 15 months after the initial surgery, the patient could barely eat or sleep, and had abandoned treatment.

Discussion

AAM is one of the rarest and perhaps most misdiagnosed types of genital stromal tumors. It is often detected incidentally during physical examinations, owing to its slow growth pattern and lack of symptoms. If they manifest, symptoms may include pelvic fullness and pressure, dysmenorrhea, and changes in bowel and bladder function [3,9-11]. No characteristic symptoms of AAM have been identified, making it difficult to diagnose this disease.

The appearance of AAM on CT is variable. It may be a well-defined, homogeneous mass that is hypodense relative to muscle, or may be predominantly cystic with solid components [12]. Characteristic appearances on MRI include hypointensity on T1-weighted images and hyperintensity on T2-weighted images. The "swirl sign" is characteristic on T2-weighted MRI [13]. With its better soft tissue resolution, MRI is more reliable for the diagnosis of AAM. MRI was performed following the first recurrence of the tumor in our patient to evaluate her condition; however, CT was used during follow-up visits owing to the patient's financial situation. Given its characteristic features, MRI should be used for delineating the extent of disease, determining eligibility for surgery and clinical follow-up examinations for recurrent tumors if a patient's financial situation allows. On gross examination, AAMs are typically soft, bulky masses. Microscopic examination shows a sparsely cellular tumor composed of pale-to-eosinophilic stroma with numerous haphazardly arranged blood vessels that stand out against a myxoid background. There is no specific immunohistochemistry marker for AAM; however, these tumors are generally positive for vimentin and desmin.

The preferred treatment for AAM is surgery, although achieving negative resection margins is challenging owing to the infiltrative nature of the tumor [14]. It is especially difficult to remove large and deep-seated tumors in the pelvis compared to the small and superficial tumors of the vulva or vagina. To assess the necessity of radical resections, researchers analyzed data from 111 patients with AAM and found no statistical differences in survival according to the time to relapse or resection margin status [2]. Although complete surgical resection should be the preferred aim, incomplete or partial resection is acceptable when significant operative morbidity is anticipated or when the preservation of fertility is required. Our patient experienced recurrence despite wide excision with negative surgical margins; she chose not to undergo a third excision after recurrence and sought chemotherapy at another hospital. Generally, adjuvant radiotherapy or chemotherapy is not a preferred route because of the low mitotic activity of AAM [3]; however, several cases of tumor recurrence have been treated with radiation therapy resulting in patients achieving relapse-free intervals of 2–3 years [15,16]. To our knowledge, ours is the first report of chemotherapy used for a patient with AAM; however, it did not provide any curative benefit.

The treatment of recurrences, however, may be challenging and require various therapeutic options to be explored. This is especially true given that no single modality has been proven to be effective [17]. AAM tends to recur after surgical excision despite being a benign and non-invasive tumor; such recurrence can occur at the original site after the initial resection [18]. Recurrence rates range from 25% to 47%, and 85% of all recurrences appear within 5 years of initial surgery [2,3,19]. It is highly recommended that patients with AAM be closely followed by using clinical and imaging tests, particularly MRI. To our knowledge, only 2 cases of metastasis from AAM have been reported in the literature [20,21], suggesting that metastasis is an exceedingly

rare event. Because recurrences have been observed 14 years after the initial diagnosis [14], proper management, combined with long-term follow-up, is necessary to identify recurrences of AAM.

Conclusion

AAM should be considered in the differential diagnosis of a painless, soft, slow-growing mass in women. Wide local excision remains the first-line treatment to date. Long-term follow-up and careful monitoring with imaging techniques are required for timely identification of the recurrent tumors and prompt resection. In our patient, chemotherapy did not provide any curative benefit.

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