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Case Report

Aggressive Angiomyxoma of the Pelviperineum: Surgical Treatment through a Perineal Incision

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ABSTRACT Aggressive angiomyxoma is a rare benign tumor of the pelvic soft tissue in women of reproductive age. The tumor is locally infiltrative and tends to recur. Herein is presented a case report of aggressive angiomyxoma that was totally excised using a transperineal approach. A 35-year-old woman had an aggressive angiomyxoma of the vulva and pelvis, with swelling of the right labium majus pudendi. Three years previously, she had undergone incomplete excision of the same type of pelviperineal mass via the transabdominal route. After a complete workup, a transperineal minimally invasive approach was used to excise the 20-cm mass filling the right side of the pelvis. Histopathologic findings were consistent with a diagnosis of aggressive angiomyxoma. Although often misdiagnosed as various other benign genital disorders, angiomyxoma usually is manifested as a soft nontender mass. After a thorough examination and full radiologic workup, a small transperineal incision may be sufficient for complete removal of the tumor. Journal of Minimally Invasive Gynecology (2011) 18, 541–544 © 2011 AAGL. All rights reserved.

Keywords: Aggressive angiomyxoma; Minimally invasive surgery; Perineum; Vulva

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Aggressive angiomyxoma is an uncommon mesenchymal neoplasm that occurs predominantly in the pelviperineal region in women of childbearing age. Its peak incidence is in the fourth decade of life, with predominance in white women [1–3]. It is a slowly growing and locally infiltrating tumor. The term "aggressive" was introduced to emphasize the locally aggressive behavior and high potential for local recurrence; it does not imply a high probability of metastasis. Radical surgery with wide margins and close follow-up is the currently accepted treatment of choice [3].

Case Report

A 35-year-old woman came to our clinic with a palpable mass on the right labium majus. Three years previously at another medical center, she had undergone incomplete surgical excision via a Pfannenstiel incision of an identical mass. Histopathologic analysis demonstrated that the pelvic mass was a benign myxofibromatous tumor. Other than 3 cesarean section deliveries, her history was unremarkable. Vulvar examination revealed a globular, rubbery, smooth mass protruding from the right labium majus and extending up to the right adnexa next to the right vaginal wall, bladder, and uterus. The overlying skin in the perineum was normal (Fig. 1).

For further investigation, magnetic resonance imaging (MRI) was performed, which revealed a heterogeneous right pelvic and vulvar mass arising from the right labium majus and extending through the right vaginal wall, adjacent to the bladder and uterus. The mass had a craniocaudal diameter of 20 cm, and was 3 to 7 cm wide (Fig. 2).

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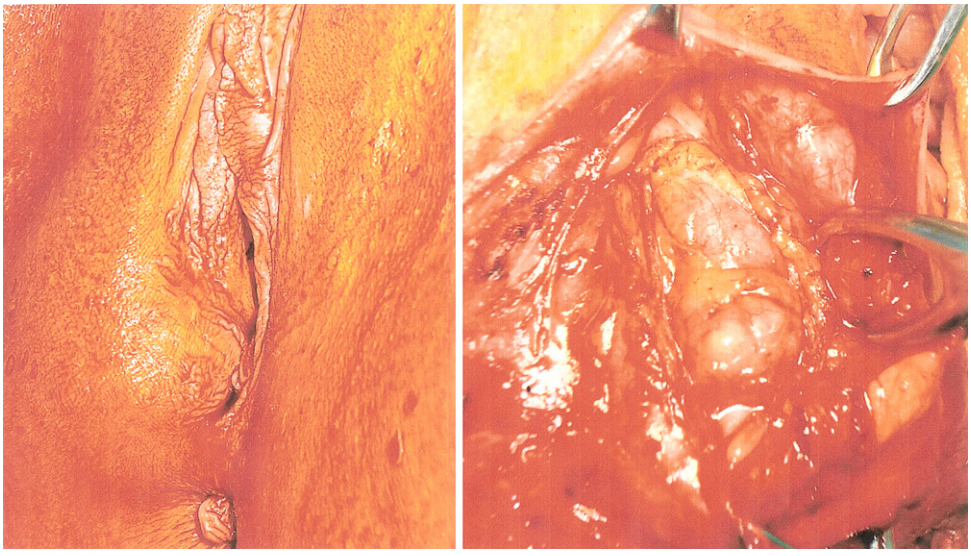
Fig. 1

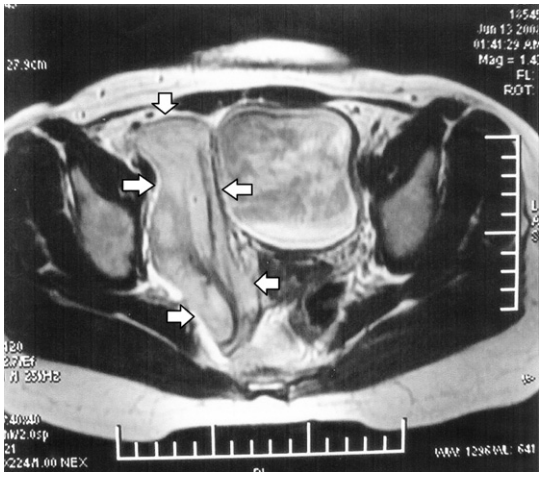
Left: Gross anatomy of the vulva shows protuberance at the level of the right labium majus with healthy overlying skin. Right: Gelatinous tumor tissue is grasped using oval forceps.



Because of the findings, the previous resection, and history of 3 cesarean section deliveries, a minimally invasive approach using transperineal surgery was performed. A 6- to 7-cm incision was made on the sulcus of the lateral border of the right labium majus pudendi, and was expanded using retractors. The gelatinous tumor tissue was readily identified and grasped using oval forceps for traction (Fig. 1). The dissection was performed primarily via sharp dissection using scissors. During the course of the surgery, the bladder was identified using a catheter and was dissected from the tumor via blunt dissection (traction-countertraction). After all tumor tissue was excised, the fossa was examined for possible residual tumor (Fig. 3). The skin incision was closed using continuous polyglactin 910 sutures (Vicryl 3/0; Ethicon, Inc., Somerville, NJ), and a suction drain was placed (Fig. 4). The postoperative course was uneventful.

Fig. 2

Magnetic resonance image of the pelvis shows tumor tissue (arrows) adjacent to the bladder.



The final pathologic diagnosis was aggressive angiomyxoma. After discussion by the multidisciplinary tumor board, the patient was given an antiestrogen, tamoxifen, for 3 months. After completion of postoperative therapy, the medication was discontinued, and repeat pelvic MRI was performed, which scan revealed neither recurrence nor residual tumor. At 18-month follow-up, both examination and MRI were negative for recurrent disease.

Discussion

First described in 1983 by Steeper and Rosai [1], aggressive angiomyxoma is an uncommon mesenchymal neoplasm that occurs predominantly in the pelviperineal region in adults. The tumor occurs primarily in women of reproductive age, which suggests that estrogen may stimulate its growth [2]. Like our patient, most patients demonstrate a slow-growing mass that is frequently the only sign. Observed accompanying symptoms include regional pain, a feeling of local pressure, and dyspareunia. Tumor size is often underestimated at physical examination. It is misdiagnosed in more than 80% of cases, and is most often mistaken for a Bartholin cyst, vulvar abscess, lipoma, Gartner duct cyst, vaginal mass or polyp, vaginal prolapse, pelvic floor hernia, or obturator or levator hernia [3–6]. Thus, imaging is extremely important to determine the extent of the