Asymptomatic aggressive angiomyxoma with a two-year follow up in a middle-aged healthy male: A case report and brief review of literature

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\textbf{ABSTRACT}

A 48-year old male found to have an asymptomatic perineal mass during hospital evaluation for urolithiasis presented two years later for follow up. He underwent imaging which showed slow growth. He subsequently underwent surgical excision of the mass. Histopathology showed it to be an aggressive angiomyxoma. He is currently being followed up on and will continue to do so due to the recurrent nature of the tumor. This case highlights the asymptomatic presentation of aggressive angiomyxomas, consideration for the differential diagnosis in males with a pelvic mass, need for research to potentially prevent recurrence, and importance of follow-up.

1. Introduction

Aggressive angiomyxomas are benign, slow-growing tumors composed of myxomatous and collagenous stroma.\textsuperscript{1} These tumors are known for their high recurrence rates with reports of around 50% and tendency for local invasion.\textsuperscript{2} Treatment is typically with surgical excision with adjuvant hormonal therapy as an option. They are rare with reports of less than 400 cases in the current literature. Theses tumors primarily present as a perineal, pelvic, or vulvar mass in reproductive-age women.\textsuperscript{1} The clinical course can vary from asymptomatic to compression of local pelvic structures causing urinary symptoms, pain, pelvic pressure, and dyspareunia. Metastasis and death is rare with just 2 cases being reported in the literature with distant metastases to the lungs. It is very rare for angiomyxomas to present in males, as few cases have been reported. Here we report a rare case of a perineal aggressive angiomyxoma in a healthy middle-aged male.

2. Case presentation

An otherwise healthy 48-year-old Caucasian male was incidentally found to have a 6.7 × 5.9 × 2.4 cm ovoid lesion in the right perineum abutting the bulbar urethra during an ED work-up for nephrolithiasis (Fig. 1). At that time, the patient was asymptomatic and recommended to undergo an MRI to further characterize the lesion.

He was lost to follow-up until 10 months later when he started to note perineal discomfort with sitting or riding a bike. On perineal examination there was a visible, non-tender without fluctuance, erythema or inguinal lymphadenopathy. The patient had no urinary symptoms. Pelvic MR revealed a circumscribed mass in the right perineum measuring 7.3 × 3 cm axially and 8 cm craniocaudally [Fig. 2]. There was mass effect on the bulbar urethra but no gross invasion. The mass was T2 bright with swirled echotexture, and an isointense T1 signal with heterogenous hyperenhancement. Vessels were seen within the lesion. No regional lymph node involvement was noted. Differential diagnosis included angiomyxoma, lipomyoma, liposarcoma, and sarcoma.

Given risk of malignancy and symptoms, patient elected to undergo surgical excision. Intraoperatively, mass was felt to be mobile. Cystoscopy and digital rectal exam were negative for invasion of urethral lumen and rectal wall, respectively. The mass was found to abut anteriorly the apex of the right corpus cavernosum, laterally the right ischial pubic ramus, medially the corporal spongiosum, posterior the peri-rectal muscle fibers, and superior the levator muscle. Patient was discharged on post-op day 1 after undergoing void trial. Final pathology revealed an 8 cm hypocellular lesion with myxoid stroma and abundant vasculature with surrounding muscular tissue, suggestive of an aggressive angiomyxoma [Fig. 3A and B]. By immunohistochemistry, stromal cells were positive for desmin and CD34 and negative for S100 and EMA. Clinical follow up is ongoing.

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2214-4420/© 2022 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).
Angiomyxoma is a rare soft tissue tumor of myofibroblastic origin, first described by Steeper and Rosai in female pelvis and perineum. In the literature, there are about 350 reported cases, with the vast majority being described in women of reproductive age. Tumor locations are mostly limited to the pelvis, perineum, and male scrotum. Extra-pelvic sites have been described, such as the kidney, liver, or orbit. There is a female predominance with female-to-male ratio of 4-6:1. Angiomyxoma is known to be benign, and metastatic disease is rare. These tumors are typically well-defined and tend to displace local structures rather than invade them. In addition, they are often misdiagnosed. Other differential diagnosis for perineal mass in men includes perineal cyst, lipoma, abscess, urothelial carcinoma, leiomyoma, leiomyosarcoma, and liposarcoma.

Angiomyxomas is a pathologic diagnosis. They are characterized on histopathology as loose stromal connective tissue containing short spindle cells on a background of collagen and myxoid with vascularization. Immunohistochemical profiles of the tumor can be helpful in differentiating angiomyxoma from other differential diagnoses such as myxoliposarcoma and histiocytoma as angiomyxomas characteristically stain positive for CD34, desmin, and vimentin. Hormonal receptors (estrogen and progesterone) can be positive as well while S-100, CD68, and actin stains are mostly negative. The tumor in our patient stained...
positive for CD34 and desmin, and negative for S100. Nevertheless, CT and MR with contrast are important diagnostic tools.

Treatment involves surgical excision with negative margins. Due to its slow-growing nature, angiomyxoma is not known to respond to chemotherapy. Radiation therapy has been reported in the palliative setting. Adjuvant hormonal therapy can be an option when tumors stain positive for ER or PR receptors. Interestingly, negative margin status is not a negative predictive factor in recurrence free survival according to this retrospective review. Long-term data on follow-up is limited. Local recurrences were reported as 47% in one case series (median follow-up unavailable; longest follow-up 14 years). Therefore, surveillance is prudent in these patients.

4. Conclusion

This report highlights a rare case of angiomyxoma in a healthy middle-aged male with a two-year asymptomatic period. Its diagnosis requires a high index of suspicion. Complete cure relies on radical excision. Long-term follow-up is warranted.

Declaration of competing interest

The authors have no conflicts of interest.

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