CASE REPORT

A Rare Case Report of Deep Aggressive Angiomyxoma Mimicking Bartholin's Cyst

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Abstract

We present a case of deep aggressive angiomyxoma (AA) of a vulval lesion. This case report is of a 32-year-old female (para 1, living 1) who presented to the gynecological outpatient department with complaints of a lump felt in her left vagina for 1 month. This was initially misdiagnosed as a Bartholin gland cyst. She underwent excision of Bartholin's gland under anesthesia. The histopathology reported a deep AA of the vulval lesion. Despite the rarity of this condition, with only 350 cases reported so far, it is crucial to include it in the range of differential diagnoses for a pelvic mass, considering the locally aggressive characteristics of this tumor and its tendency to recur.

Keywords: Angiomyxoma, Bartholin's cyst, Case report, Recurrence, Vulval lesion.

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Introduction

Angiomyxoma represents an infrequent benign mesenchymal tumor characterized by a gradual growth pattern, typically manifesting as a low-grade neoplasm predominantly found in the vulvovaginal (perineal) area. This tumor exhibits a propensity for recurrence and infiltration into adjacent skeletal muscle and adipose tissue.[1] This uncommon soft tissue tumor was previously known as aggressive angiomyxoma (AA) and was initially identified and detailed by Steeper and Rosai in their case series in 1983.[2]

In 2003, the World Health Organization reclassified the term to designate it as deep angiomyxoma.[3] This tumor is notably uncommon, with fewer than 350 reported cases to date.[4] Aggressive angiomyxoma predominantly affects women during their reproductive years, displaying a heightened occurrence typically observed between the third and fifth decades of life.

This lesion is derived from myxoid cells of connective tissue and typically manifests as a painless mass in the vulvoperineal region. The term "aggressive" is used to describe its pronounced inclination for local invasion, often infiltrating the perivaginal and pararectal tissues. Despite being mostly benign in nature, the tumor has frequent local recurrences, with rates ranging from 30 to 72%.[5] However, the metastatic potential for the lesion is relatively low.

Misdiagnosis of AA is often reported, and it is frequently misidentified as a Bartholin gland cyst, vulvar abscess, lipoma, or hernia, primarily due to a considerable part of the tumor being hidden within the depths of the soft tissues in the pelvic region. Sato et al. reported cases of AA originating beyond the pelvic region and this was managed by extensive surgical resection.[6] We discuss an interesting case of AA in a 32-year-old female which was initially diagnosed as Bartholin cyst and then correctly diagnosed via histopathology.

Case Description

Mrs X, 32 years old, attended our gynecology clinic with complaints of a lump felt in the left side of the vagina for 1 month. She also complained of feeling pain and discomfort while walking. She felt that the lump had increased in size compared to how it was previously. She also complained of vulval itching associated with vaginal discharge.

Mrs X was Para 1 with one 5-year-old child who was delivered via normal vaginal delivery. Her past medical and surgical history were unremarkable. On local examination, a non-tender lump was seen in the lower end of the left labia majora and measured about 3 cm in size. An impression of left Bartholin's cyst was made and she was counseled for marsupialization of the cyst.

She underwent excision of the Bartholin's gland under anesthesia. A linear incision was made on the inner aspect of the labia minora. The incision was deepened but no secretion was drained. A spongy mass was identified. A plane of cleavage was obtained and the mass was completely enucleated and sent for histopathology. During enucleation, the base was very vascular, and hemostasis was obtained with deep sutures. The differential diagnosis was Bartholin's gland/Gartner cyst.

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The histopathology report identified a deep AA of the vulval lesion. Sections revealed an unencapsulated hypocellular mesenchymal lesion characterized by the proliferation of spindle to stellate cells. These cells displayed delicate cytoplasmic processes, bland chromatin, and small nucleoli within a myxoid stroma. The stroma contained scattered delicate collagen fibers, along with conspicuous haphazard dilated capillaries and perivascular stromal smooth muscle bundles. Increased mitotic activity, significant cytologic atypia, or necrosis were not seen. On evaluation with immunohistochemistry, the lesion cells were positive for estrogen receptors positive (ER) and progesterone receptors positive (PR), while perivascular stromal smooth muscle bundles were highlighted by desmin. The lesion cells were negative for CD34 and S100. The immunohistochemical studies were performed with positive and negative controls, showing appropriate reactivity (block A3).

She presented after 10 days of surgery with complaints of pelvic and perineal pain for 1 week, associated with chills and rigors for the past 2 days. On examination, a 3–4 cm swelling was observed in the left lower third of the left labia minora. Oozing of dark blood was noted from the vulvar incision at the site of the previous surgery. A diagnosis of vulvar hematoma was made, and she was scheduled for evacuation of the hematoma. This was done under anesthesia with subsequent suturing of the vulvar incision. A 1 cm piece of bulging vulvar tissue at the incision site was excised and sent for histopathology. The histopathology report reaffirmed the diagnosis of deep AA. The patient was discharged in stable condition with a referral to the oncology department. Her case was discussed in the multi-disciplinary team meeting, and it was decided to do an MRI of the perineum 4 weeks after discharge.

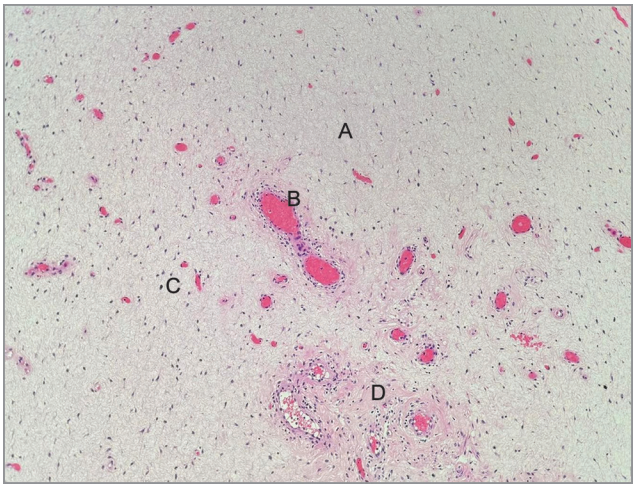

Fig. 1: Histopathology slide of the patient showing deep aggressive angiomyxoma. A, Prominent myxoidstroma; B, Vessels of varying caliber; C, Spindle cells; D, Perivascular smooth muscle and collagen cuffing

Discussion

Angiomyxoma is a non-malignant neoplastic condition arising from mesenchymal tissues, characterized by significant local invasiveness and a propensity for frequent recurrence.[7] Typically exhibiting a pink hue, angiomyxoma presents with a rubbery texture and a glossy surface. Due to its typical location, it is frequently mistaken for various conditions such as Bartholin's gland cyst, labial cyst, Gartner duct cyst, vaginal cyst, abscess, leiomyoma, fibroepithelial polyp, sarcoma, lipoma, or conditions like inguinal/femoral hernia, vaginal prolapse, and gynecological malignancies.

The reported rates of misdiagnosis can be as high as 82%.[8] Djusad et al. reported a case report in 2021 of a deep AA which was initially diagnosed as a Gartner duct cyst, with the correct diagnosis being established with histopathological examination.[8] Another case report by Devi et al. similarly reported to have misdiagnosed a case of AA as a Bartholin's cyst. The correct diagnosis was ultimately determined after histopathological examination.[9] The prevalence of AA in the general population remains uncertain due to its rarity, complicating both management and counseling. Typically, AA tumors are characterized by substantial size, often exceeding 10 cm in their largest diameter. These tumors display a visibly lobulated structure on a macroscopic level and can adhere to the adjacent soft tissue. Under microscopic examination, one can observe spindle or stellate-shaped cells entwined within a loose matrix containing wavy collagen and edema. The cellularity tends to range from moderate to low, and infiltration into fat, muscle, and nerves is discernible. A distinctive feature of AA is the presence of vessels of varying calibers scattered haphazardly throughout the tumor parenchyma, while mitotic figures are infrequent (Fig. 1). On immunohistochemical examination, desmin and smooth muscle progesterone receptor are typically positive, and some tumors may exhibit positivity for CD34, with consistently negative expression of S100. Most AA specimens express estrogen and progesterone receptors, suggesting a hormone-dependent growth. Consequently, treatment with GnRH agonists has been administered to patients with AA, and there are documented cases reporting significant responses to such treatment.[10]

Utilizing imaging techniques such as USG, CT, and MRI is effective in diagnosing angiomyxoma. MRI, particularly on T2-weighted images, reveals high signal intensity and a swirling appearance consistent with the tumor's high water content and myxoid composition, mirroring findings in CT. In CT imaging, angiomyxoma might manifest as hypodense or exhibit a similar attenuation to the surrounding skeletal muscle, displaying a combination of cystic and solid components. A comprehensive evaluation involving transabdominal, transperineal, and transvaginal ultrasound examinations can contribute to a thorough assessment, characterizing the lesion as a hypoechoic and heterogeneous mass.[11]

The rarity of this condition poses challenges for making a preoperative diagnosis. The management involves the cyst excision. This includes a longitudinal incision in the vaginal mucosa, followed by a sharp dissection to separate the cyst wall until reaching its base. The base of the cyst is then cut, and the vaginal mucosa is sutured. Surgical excision with wide margins remains the preferred treatment option.[12] Complete removal through surgery is challenging due to infiltration into the surrounding tissue, leading to common recurrence in the ischiorectal and retroperitoneal spaces. The potential for recurrence arises from both deliberate and unintentional incomplete removal, as identifying the tumor borders can be challenging, even with what seems to be adequate resection. Additionally, as per a retrospective assessment carried out by Chan et al., individuals with positive margins demonstrated a similar probability of recurrence when compared to those with negative margins.[13]

Surgeons may contemplate addressing residual disease through hormone-modulating therapy or alternatively choose close observation. The efficacy of this approach has been demonstrated

over extended follow-up periods, showing no subsequent recurrence of the tumor. GnRH agonists contribute to tumor size reduction before excision and can effectively address recurrence, given the presence of estrogen receptors.[14] Additionally, Raloxifen and Tamoxifen serve as neo-adjuvant therapies for residual or recurrent tumor growth. Strategies like preoperative embolization or external beam irradiation can help diminish the likelihood of recurrence. However, there is no single modality that has proven more advantageous than others. To monitor for local recurrence and invasion, and critical sources of morbidity, a close follow-up is advisable for at least 2 years. Long-term surveillance involves the use of MRI, recognized as the most effective imaging modality for detecting recurrences.[15]

In our case, a multi-disciplinary meeting was held, and it was decided to perform an MRI of the perineum 4 weeks after discharge.

Conclusion

This case report describes the initial misdiagnosis of an AA as a Bartholin's gland cyst. The infrequency of AA poses challenges in making an accurate preoperative diagnosis. Conducting both biopsy and imaging procedures contributes to achieving an accurate preoperative diagnosis. Despite its rarity, AA should be included in the list of potential differential diagnoses for vulvar tumors. Finally, long-term follow-up with clinical examination is imperative for proper monitoring and management, considering the high recurrence rate of this condition.

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