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Diffuse Aggressive Angiomyxoma of the Vulva: A Case Report

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Summary

Aggressive angiomyxoma is a mesenchymal tumor that is frequently found in women of reproductive age mostly appearing in the vulva, perineal region, head, neck, trunk, and lower limbs. A 33-year-old multiparous Somali woman presented to the outpatient department of Dr Sumait Hospital, with a complaint of swelling around her vulva. The patient reported that the mass had progressively grown for the past 8 years and that she experienced stigma from her previous spouses. Her past medical history and socioeconomic status were unremarkable. During the physical examination, a well-defined pedunculated mass measuring 14 cm × 8 cm, originating from the labia majora, was noted. The mass was pinkish, non-tender, non-reducible, and had a soft consistency. Some areas of ulceration measuring 2 cm × 2 cm, covered with necrotic granulation tissues, were also noticed. With the patient's written informed consent, a biopsy was performed, and the histopathology results revealed aggressive angiomyxoma. Following confirmation of the diagnosis, the mass was surgically excised under sterile conditions and spinal anesthesia. Aggressive angiomyxoma is an important consideration when evaluating vulvovaginal growth in females. Given its tendency for local recurrences, it is vital to establish an accurate diagnosis.

Keywords: Aggressive angiomyxomas, Case report, Labia majora, Mesenchymal tumor, Somalia

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Introduction

Aggressive angiomyxomas are rare mesenchymal tumors that primarily affect women of childbearing age, with most cases occurring around the fourth or fifth decade of life (1, 2). Patients are often asymptomatic at the time of diagnosis, and the presence of perineal or vulvar masses is typically discovered incidentally during physical examinations or radiological imaging (2). Although aggressive angiomyxomas are uncommon, their ultrasound, computed tomography (CT) scan, and magnetic resonance imaging (MRI) characteristics exhibit similarities. These tumors consistently display a distinct "swirled" appearance on MRI, characterized by

relatively low intensity and internal stranding on both T1- and T2-weighted images within the perineal and/or pelvic regions (3, 4).

This case report aims to familiarize physicians with the clinical and histopathological features of aggressive angiomyxoma.

Although the likelihood of metastasis is minimal according to numerous studies, the rate at which the condition is misdiagnosed varies from 70% to 100%, leading to inadequate and delayed treatment that exacerbates patient suffering. Although extensive local excision with negative margins and long-term monitoring remains the recommended approach, it often results in significant morbidity. In addition, adjuvant treatment with gonadotropin-releasing hormone (GnRH) agonists have shown promising results in terms of preventing recurrence (5).

Case presentation

A 33-year-old woman, with an incomplete medical history, discovered a swelling on the right side of her vulva about 8 years ago. The patient reported that over time, the swelling had progressively increased in size and now presented as a substantial, pinkish, pedunculated mass (Figure 1A). Further enquiry revealed that her medical and family history was unremarkable.

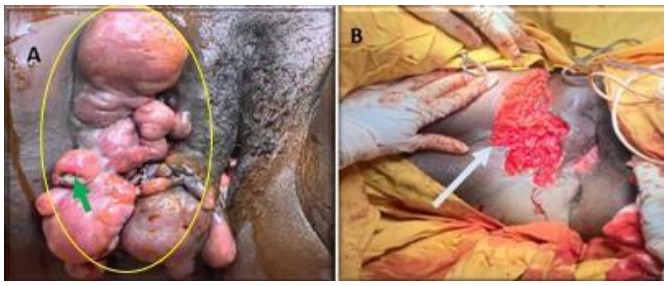

Figure 1: Lesions on presentation and after surgery. (A) Pedunculated tumor (circled with green arrow showing ulceration) arising from the right vulva with a pinkish, and multi-lobular appearance. (B) Postoperative picture of the area surrounding the vulva after resection of the angiomyxoma (arrow).

Upon physical examination, she had an average body build and showed no signs of fever, abnormal skin pigmentation, jaundice, finger clubbing, or lymphadenopathy. Further physical examination revealed a well-defined, multi-lobulated pedunculated mass measuring 14 cm × 8 cm on the right labia majora of the patient's vulva, which also had small ulcerations. Palpation revealed that the mass was non-tender, non-reducible, and exhibited a soft consistency. Additionally, there were areas of ulceration measuring 2 cm × 2 cm, covered with necrotic granulation tissue. Hematological investigations, including a complete blood count, showed all parameters to be within the normal range, with a hemoglobin level of 13 g/dL. Other laboratory tests, such as the basic metabolic panel, renal panel, and hepatic function tests, yielded unremarkable results. However, due to financial constraints, an MRI could not be performed.

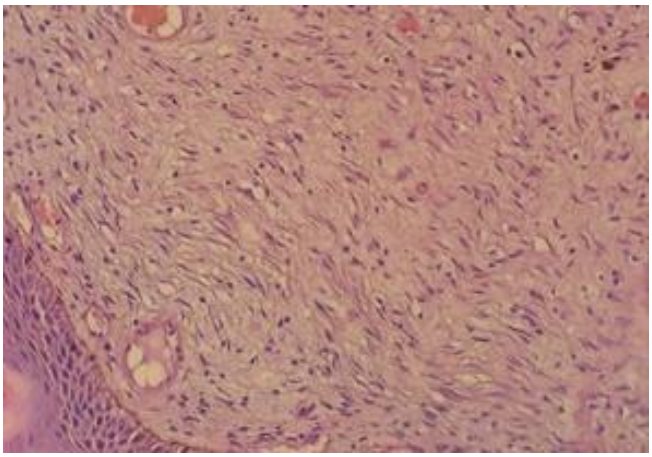

Figure 2: Histological section of the polypoid tissue showing myxoid stroma.

Following informed written consent, an excisional biopsy sample was taken, and the histopathology results confirmed the diagnosis of aggressive angiomyxoma. The histopathological examination revealed spindle and stellate-shaped cells in a myxoid matrix suggestive of aggressive angiomyxoma with tumor-free margins (Figure 2). One week later, after receiving the histopathology results, a wide local excision of the mass and surrounding margins was performed under local anesthesia (Figure 1B).

Two weeks after the surgical excision and post-surgical management of the case, the sutures were removed, and the wound was completely healed with significant scar

formation (Figure 3). As Somalia has no oncology center, getting chemo and radiotherapy treatments is quite expensive and the family could not afford the cost.

Figure 3: Two weeks postoperative picture of vulva after resection of angiomyxoma.

Discussion

Angiomyxomas can manifest as either aggressive (cutaneous myxoma) or superficial types. Superficial angiomyxomas are more commonly observed in middle-aged individuals and often present as a single large nodular mass or polypoidal growth with multiple lobules, primarily appearing on the neck, head, and lower limbs (6). They are frequently associated with Carney's complex and are prone to being misdiagnosed as skin tags, cysts, or neurofibromas. Although rare, aggressive angiomyxomas can also occur in perimenopausal women and children. However, they are predominantly documented in women of reproductive age (6). This could be attributed to the tumor's hormone sensitivity, as evidenced by cases of rapid growth during pregnancy (6). Estrogen and progesterone promote tumor growth. Typically, aggressive angiomyxomas are observed in the pelvic or perineal region, although uncommon locations include the lung, laryngeal area, and occasionally the liver and orbit (3). In male patients, the inguinal region and scrotum are the commonly affected sites, and there is a higher recurrence ratio in females compared with males (6:1) (7). The exact cause of aggressive angiomyxoma remains unclear. However, some researchers suggest that it may originate from proliferative mesenchymal cells or multipotent perivascular stem cells (8).

The clinical presentation of the present case was similar to cases of aggressive angiomyxoma reported in the literature, which are usually characterized by large pedunculated, and painless polypoidal mass around the vulva. Patients with aggressive angiomyxomas are asymptomatic and came with nonspecific symptoms such as of feeling local pressure in the vulva area, as well as the gluteal and suprapubic regions with a typical dull aching pain, and urinary retention. The size of most angiomyxoma varies between 1 and 60 cm. The severity of the condition is frequently underrated due to its local nature following physical examination; however, aggressive angiomyxomas have been reported to be widely involved with deeper tissues around the pelvis.

An aggressive angiomyxoma can be differentiated from other neoplasms by its characteristic vascularity, distinguishing it from myxoma, myxoid sarcomas, angiofibroblastoma, vulva hypertrophy with lymphedema, lipoma, and sarcoma. Surgical excision is considered as the preferred treatment for aggressive angiomyxoma (9, 10). Additional treatment options include hormonal therapies such as raloxifene, tamoxifen, and GnRH analogs, which can help shrink the tumor and prevent recurrence (9, 10). Owing to the low tendency of this tumor to metastasize, chemotherapy and radiation therapy are not commonly recommended (11). Embolization is contraindicated due to the tumor's high vascularity (12). It is important to note that this was the first time the case was managed, so the extent of the tumor and the possibility of recurrence cannot be definitively determined. The patient did not receive hormonal therapy due to financial constraints and the unavailability of medication. However, she was advised to promptly report any signs of recurrence to the hospital.

Although aggressive angiomyxomas are histologically benign, they have a high local recurrence rate, which justifies the term "aggressive" to describe their infiltrative nature and frequency of return (13). In a similar case study, 34 out of 73 cases managed showed a recurrence rate of 47% (6). While rare, a few cases of

aggressive angiomyxoma have been reported to metastasize, leading to fatalities (8). Historically, aggressive angiomyxoma was believed to be a nonmetastasizing tumor. However, there have been rare reports in the literature documenting cases of metastasis. Siassi et al. reported a fatal case in which the tumor metastasized to multiple organs, including the peritoneum, lungs, and lymph nodes (13). Another case involved a 34-year-old woman who experienced local recurrences following the initial resection of an aggressive angiomyxoma, eventually succumbing to multiple lung metastases (11). While there are currently no evidence-based guidelines for post-surgery management of aggressive angiomyxoma, due to the high rate of local recurrences and the potential for metastasis, it is recommended that patients undergo long-term follow-up for up to 15 years after the initial excision.

Conclusion

Aggressive angiomyxoma is an uncommon and locally invasive tumor that should be considered as a potential differential diagnosis in cases involving growth in the vulvovaginal region, perineum, or pelvis. Given its propensity for local recurrences, prompt and accurate diagnosis, followed by surgical excision and adjuvant therapy, are crucial for effective management of affected patients.

Ethical consideration

Informed consent was acquired from the patient for publication of the case report.

Author contributions

M.A. conducted the surgery; F.I. and M.A. carried out the postoperative care; M.A.K. prepared the histopathology report; A.M. was responsible for writing the original draft preparation; and B.G. contributed to review writing and editing. All authors have read and agreed to the published version of the manuscript.

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