CASE REPORT

Aggressive angiomyxoma of pelvis: Diagnostic challenge

Vijayalaxmi S Patil[1], Neelamma Patil[2], Rahul Kanungo[1]*

[1]Department of Pathology, [2]Department of Obstetricis and Gynaecology, BLDE (Deemed to be University) Shri B. M. Patil Medical College, Hospital & Research Centre, Vijayapura-586103 (Karnataka), India

Abstract

Aggressive Angiomyxoma (AA) is a rare, benign, and slow-growing mesenchymal tumor that affects the female pelvis and perineum. It appears as an infiltrative tumor with a high likelihood of local recurrence (up to 71%), which is why it is called aggressive. In this case, a 35-year-old female presented with heavy menstrual bleeding for four months and abdominal pain for one month. Clinical examination revealed the presence of a palpable, uniform, non-mobile mass over the uterus. Ultrasonography revealed fibroid with cystic change. Surgical excision of the mass was performed and its histopathological examination established the diagnosis as aggressive angiomyxoma. The diagnosis of AA is difficult due to its nonspecific clinical and radiological findings. This case report reveals the importance of histopathology in the diagnosis of aggressive angiomyxoma.

Keywords: Aggressive, Angiomyxoma, Pelvic Mass

Introduction

Aggressive Angiomyxoma (AA) is a mesenchymal tumor that is rare, benign, and slow- growing, and it primarily affects the lower genital tract of women during their reproductive years [1]. Its incidence is six times higher in women than in men, and it is commonly found in the deep soft tissues of the vulvovaginal region, pelvis, and perineum in women, and the analogous inguinoscrotal region in men [2-3]. The symptoms of AA may be nonspecific, including pelvic fullness, vulvar or perineal pain, dyspareunia, or no symptoms at all. While the tumor may cause a mass effect, it rarely invades the vagina, rectum, or urethra [4]. AA is so named because of its myxomatous stroma and vascular component. It is considered an infiltrative, slow-growing, and benign-looking tumor with a high risk of local recurrence (up to 71%), which may occur many years after the initial diagnosis. It is locally aggressive, but the probability of metastasis is low [5]. The diagnosis of AA can be challenging due to its nonspecific clinical and radiological findings, and histopathological examination is typically necessary to establish a definitive diagnosis [6]. This report describes a case of AA located on the posterior wall of the uterus.

Case Report

A 35 year old female presented with heavy menstrual bleeding for 4 months and with pain in abdomen for 1 month. On examination, the uterus was found to be of 22-24 weeks size of a gravid uterus, along with the presence of a palpable, uniform, non-mobile mass. Ultrasonography of the abdomen and pelvis was performed and it revealed a large hetero-echoic lesion along the posterior aspect of the wall and fundus of the uterus, having

central vascularity and showing peripheral cystic changes suggestive of fibroid with cystic change. Surgical excision of the mass was done. Gross examination of the specimen revealed an encapsulated mass measuring 15 × 12 × 6.5 cm with bosselated external surface. Cut section of the mass showed presence of light green colored gelatinous areas along with microcysts filled with pale myxoid material (Figure 1). Focal solid grey white to pale brown areas were also noted. Multiple sections (upto 12) from various representative areas of the mass were given. Histopathological examination of the sections from the mass revealed hypocellular tumor tissue comprised of loose myxoid stroma with interspersed stellate and spindle shaped cells (Figure 3). Many thick and thin blood vessels (Figure 2), congested blood vessels with extravasation of RBCs, blood vessels with perivascular hyalinization, few scattered lymphocytes and mast cells in the stroma were also noted. Based on the above mentioned gross and microscopic features, a diagnosis of AA was made.

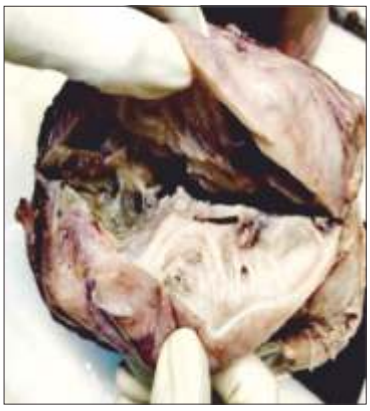

Figure 1: Cut section of the excised mass: light green colored gelatinous areas along with myxoid areas

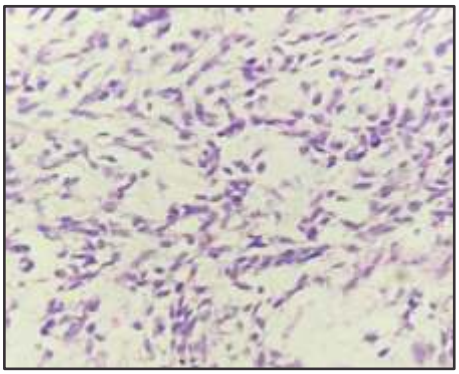

Figure 2: Hypocellular loose myxoid stroma with interspersed stellate and spindle-shaped cells

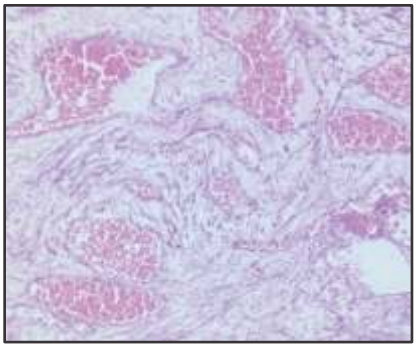

Figure 3: Many congested and thickened blood vessels (H&E, 40×)

Discussion

AA is a rare mesenchymal tumour that typically develops in the soft tissue of the pelvis and perineum in women of reproductive age. Steeper and Rosai initially characterised this tumour in their classic case series in 1983 [7]. Angiomyxomas are divided into two types, superficial angiomyxoma and deep or AA. Superficial angiomyxomas are commonly seen in males on the trunk and neck as a single nodule or a polypoidal cutaneous lesion. Superficial angiomyxomas are comprised of scattered, small to medium sized, thin-walled blood vessels in a myxoid stroma. On the other hand, AA are commonly seen in women (male-to-

female ratio 1:6) with a peak incidence in 3rd to 5th decades, located typically in pelvic and perineal region and characterized histologically by myxoid stroma and prominent vasculature of large caliber blood vessels [8]. The tumor is known to locally infiltrate the tissues and has been frequently associated with recurrence and therefore termed "aggressive". AA is a rare tumor with a total of 350 cases being reported till date, notably involving pelvis or vulvar region in most of the reported cases. Histopathology is considered the gold standard for diagnosing AA because the tumor is often misdiagnosed clinically based on its non-specific symptoms and rare occurrence [9].

The tumor is hypocellular, composed of uniform spindle/stellate cells sparsely distributed in a myxoid stroma, which is rich in an oedematous mucoid and collagen matrix, and has many thin-walled and thick-walled blood vessels. No mitotic activity or nuclear atypia is noted. There is no specific immunohistochemical marker for AA as these tumors can show variable expression of vimentin, desmin, CD34, oestrogen receptor, and progesterone receptor. The tumors can also show low proliferation index (low Ki67) and S-100 is not generally expressed [1].

The lesions namely angiomyofibroblastoma, fibro-epithelial stromal polyp, superficial cervicovaginal myofibroblastoma, cellular angiofibroma, are also known to occur in lower genital tract and they have to be differentiated on microscopy from AA. All the above mentioned lesions are superficially located and are cellular characterized by the presence of fibrous stroma while AA is deep seated and hypocellular characterized by presence of myxoid stroma [1]. The primary approach to treating AA is through surgical removal of the tumor. Excisional resection of AA is often very challenging due to the tumor's aggressive nature and its ability to invade adjacent organs [10]. In recent years, hormonal therapy using gonadotropin releasing hormone agonists and selective estrogen receptor modulators has emerged as a newer approach to treating AA. These therapies can reduce tumor volume before surgery, prevent recurrence of the tumor, and treat any residual or recurrent tumor. Radiation therapy and chemo-therapy have no definitive role in treating AA, although some cases have reported reduction in tumor size with radiation therapy [4]. The recurrence rate for AA ranges from 35-72%, with relapse typically occurring within the first 5 years after surgery. Long-term follow-up is necessary for patients after surgery [1]. Although many options have been tried for the treatment of recurrences that have varying rates of success, still there is no single modality that is clearly beneficial over other modalities [8].

Conclusion

AA is an extremely rare, locally aggressive mesenchymal tumour occurring predominantly in vulvovaginal, pelvis and perineum of premenopausal women. Histopathological examination is the preferred standard for diagnosing AA. It should be considered as a differential diagnosis especially when a patient presents with a mass, particularly in the pelvis or perineum

References

1. Lin XM, Wang L, Wang Q. Aggressive angiomyxoma of pelvis: A case report and literature review. *Medicine (Baltimore)* 2022; 101(46):e31617.
2. Raptin C, Lucot JP, Bassil A, Poncelet E, Prolongeau JF, Phalippou J. Aggressive angiomyxoma of the perineal region. *SAGE Open Med Case Rep* 2019; 7:2050313X19843391.
3. Srivastava V, Jha PK, Verma AK, Ansari MA. Vulvar aggressive angiomyxoma: A surgical challenge. *BMJ Case Rep* 2021;14:e240687
4. Akram H, Tran D, Rehman R, Al-Wahab Z. Aggressive angiomyxoma of left buttock. *BMJ Case Rep* 2021; 14(6):e241550.
5. Djusad S, Sari YM, Tjahjadi H. Deep (aggressive) angiomyxoma of the vagina misdiagnosed as Gartner cyst: A case report. *Int J Surg Case Rep* 2021; 83: 105948.
6. Alosaimi AM, Al-Jifree HM, Alharbi SY, Algethami AS. Aggressive Angiomyxoma of the Posterior Wall of the Uterus. *Cureus* 2020; 12(12):e12023.
7. Peterknecht E, Agerbak E, Mohamedahmed A, Stonelake S, Kulkarni K, Peravali R, *et al*. Aggressive angiomyxoma of the ischioanal fossa in a post-menopausal woman. *Ann R Coll Surg Engl* 2021; 103(2):e59-e64.
8. Revanna R, Ramalingappa P, Kumar DT. Aggressive angiomyxoma of uterine corpus. *Int J Reprod Contracept Obstet Gynecol* 2022; 11(7): 2023-2026.
9. York D, Saikumar S, Patel P, Edwards C, Garcia G, Naqvi H. A Paraurethral Aggressive (Deep) Angiomyxoma. *Case Rep Obstet Gynecol* 2022; 2022: 5604460.
10. Akhavan S, Nikfar S, Behboudi B, Malek M, Saffar H, Zamani N. Aggressive angiomyxoma of the pelvis surgical management in a case with delayed diagnosis. *Int J Surg Case Rep* 2021; 81:105756.

Author for Correspondence:
Dr. Vijayalaxmi S Patil, Department of Pathology, BLDE (Deemed to be University) Shri B. M. Patil Medical College, Hospital & Research Centre, Vijayapura-586103, Karnataka
Email: vijayalaxmi.patil@bldedu.ac.in
Cell: 9845417697

How to cite this article:
Patil V, Patil N, Kanungo R. Aggressive angiomyxoma of pelvis: Diagnostic challenge. *J Krishna Inst Med Sci Univ* 2023; 12(3):122-125

Submitted: *04-Mar-2023* ***Accepted:*** *04-May-2023* ***Published:*** *01-July-2023*