Case Report ISSN (O):2395-2822; ISSN (P):2395-2814

Aggressive Angiomyxoma of Vagina: A Rare Case Report.

Shilp Rani[1], Ninder Kumar[2], Prabhjot[1], Simrat Jit Kaur[1], Ramesh Kumar Kundal[3]

[1]Junior Resident, Department of Pathology, Government Medical College, Patiala.
[2]Assistant Professor, Department of Pathology, Government Medical College, Patiala.
[3]Professor & Head, Department of Pathology, Government Medical College, Patiala.

Received: November 2017
Accepted: November 2017

ABSTRACT

Aggressive angiomyxoma is a rare, locally invasive mesenchymal tumor predominantly presenting in women of reproductive age and also having a moderate-to-high risk for local relapse. Hence, it needs to be differentiated from other mesenchymal tumors occurring in this region. We present here a case of a 47 Year old female, with chief complaints of prolapsed uterus since 6 months. During clinical examination, posterior vaginal wall swelling was also identified. Total Abdominal Hysterectomy with Bilateral Salpingoophrectomy was done along with removal of posterior vaginal wall swelling. On histopathology, diagnosis of aggressive angiomyxoma was made. We report this case because of its rarity.

Keywords: Aggressive angiomyxoma, pelvis.

INTRODUCTION

Aggressive angiomyxomas are rare locally aggressive myxoid mesenchymal tumors that occur predominantly in females of reproductive age with female to male ratio 6:1, and peak incidence in the 4th to 5th decades of life.[1,2] It was first described by Steeper and Rosai in 1983.[3] It was named aggressive due to its characteristically slow and insidious growth as well as carrying a moderate-to-high risk of local relapse. It is of unknown etiology and usually affects vulva, perineal region, buttocks, or pelvis of women in reproductive age. Aggressive angiomyxomas exhibit typical patterns on ultrasound, CT, and MR imaging: tumors nearly always occur within the perineal and/or pelvic region, and on MRI characteristically demonstrate a "swirled" appearance of relatively low-intensity internal stranding on both T1- and T2-weighted images.[4-6] Because of its rarity, it is often initially misdiagnosed, frequently as a gynecological malignancy.

CASE REPORT

A 47 Year old female, resident of Patiala, presented in gynae OPD with chief complaints of prolapsed uterus since 6 months. During physical examination, posterior vaginal wall swelling measuring 4 x 3 cm was also identified. Swelling was firm and non tender. Total Abdominal Hysterectomy with Bilateral Salpingoophrectomy was done along with removal of posterior vaginal wall swelling. The specimen was sent for histopathological examination in the department of pathology, Government Medical College Patiala.

On gross examination globular soft tissue piece, tan-gray in color, measuring 5x4x3 cm was received. Cut section was soft to firm in consistency and cut surface was homogenous and gelatinous.

Figure 1: Gross: globular encapsulated soft tissue piece.

On microscopic examination monotonous and hypocellular stroma composed of small spindled and stellate fibroblasts was seen. Stroma was myxoid with collagen fibres and prominent dilated thick walled vessels, some of them showing hyalinization. On IHC, it was weakly positive for ER.

Name & Address of Corresponding Author
Dr. Shilp Rani
Junior Resident,
Department of Pathology,
Government Medical College,
Sangrur Road, Patiala – 147001.

Figure 2: Cut surface : homogenous and gelatinous.

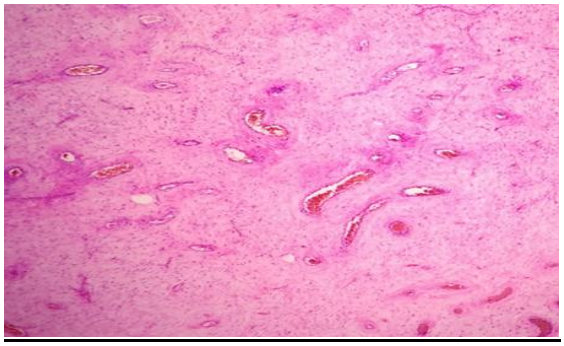

Figure 3: Photomicrograph of hypocellular stroma (H&E, ×40).

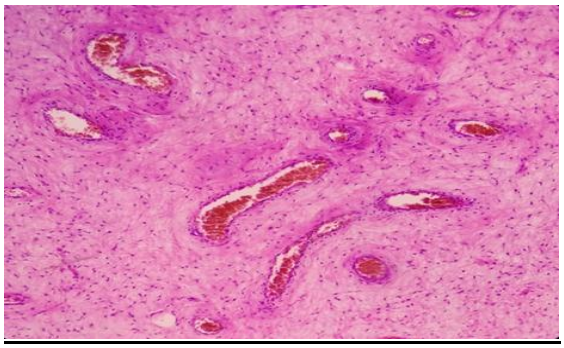

Figure 4: Photomicrograph showing prominent dilated thick walled vessels, some of them showing hyalinization (H&E, ×100).

Figure 5: Photomicrograph showing fibrosis (H&E, ×40).

DISCUSSION & CONCLUSION

AA is a hormonally responsive tumor which is often positive for ER and PR. It is believed to arise from specialized mesenchymal cells of the pelvic or perineal region. Sometimes suggest that AA arises from multipotent perivascular progenitor cells because it often displays variable myofibroblastic and fibroblastic features.[7] Microscopically, the tumor consists of spindle and stellate-shaped cells in a myxoid matrix expressing vimentin, desmin, and smooth muscle antigen (SMA) but the cells are negative for S-100.[8]

The main differential diagnosis for aggressive angiomyxomas is angiomyofibroblastoma.[9] The tumor's size is a useful differentiating factor: angiomyofibroblastomas tend to be small and affect only the superficial vulva and vagina, whereas aggressive angiomyxomas are often large masses involving the deep tissues planes at the time of diagnosis.[10] Myxomas and myxoid liposarcomas also come under differential. However, myxomas are located intramuscularly, whereas aggressive angiomxyomas may abut, but do not invade, the pelvic and/or perineal musculature. Myxoid liposarcomas more commonly occur within the lower extremities, demonstrate lacy or linear internal fat, and homogenously enhance while Aggressive angiomyxomas lack significant internal fat and enhance heterogeneously.[10]

REFERENCES

1. Sutton B.J., Laudadio J. Aggressive angiomyxoma. Arch Pathol Lab Med. 2012;136:217–221.
2. Surabhi V.R., Garg N., Furmovitz M., Bhosale P., Prasad S.R., Meis J.M. Aggressive angiomyxomas: a comprehensive imaging review with clinical and histopathologic correlation. AJR Am J Roentgenol. 2014;202:1171–1178.
3. Steeper TA, Rosai J. Aggressive angiomyxoma of the female pelvis and perineum. Report of nine cases of a distinctive type of gynecologic soft-tissue neoplasm. Am J Surg Pathol. 1983;7:463–75.
4. Kura M.M., Jindal S.R., Khemani U.N. Aggressive angiomyxoma of the vulva: an uncommon entity. Indian Dermatol Online J. 2012;3:128–130.
5. Outwater E.K., Marchetto B.E., Wagner B.J., Siegelman E.S. Aggressive angiomyxoma: findings on CT and MR imaging. AJR Am J Roentgenol. 1999;172:435–438.
6. Srinivasan S., Krishnan V., Ali S.Z. "Swirl sign" of aggressive angiomyxoma-a lesser known diagnostic sign. Clin Imaging. 2014;38:751–754.
7. Alameda F, Munne A, Baro T, Iglesias M, Condom E, Lloreta-Trull J, et al. Vulvar angiomyxoma, aggressive angiomyxoma, and angiomyofibroblastoma: an immunohistochemical and ultrastructural study. Ultrastruct Pathol. 2006;30:193–205
8. Graadt van Roggen JF, Hogendoorn PC, Fletcher CD. Myxoid tumours of soft tissue. Histopathology. 1999;35:291–312.
9. Aggressive angiomyxoma of the vulva extending into the pelvis: report of two cases. J Obstet Gynaecol Res. 2005;31:310–313.
10. Kim H.M., Pyo J.E., Cho N.H., Kim N.K., Kim M. MR imaging of aggressive angiomyxoma of the female pelvis: case report. J Korean Soc Magnetic Res Med. 2008;12:206–210.

How to cite this article: Rani S, Kumar N, Prabhjot, Kaur SJ, Kundal RK. Aggressive Angiomyxoma of Vagina: A Rare Case Report. Ann. Int. Med. Den. Res. 2018; 4(1):PT12-PT14.

Source of Support: Nil, **Conflict of Interest:** None declared