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Case Study

Aggressive Angiomyxoma of the Vulva: A Case Report

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ABSTRACT

Aggressive angiomyxoma is an uncommon and distinctive, mesenchymal tumor with a predilection for the pelvis and perineal regions in females and less frequently in males. Steeper and Rosai first described its histologic characteristics and its tendency to local infiltration and recurrence in 1983. About 90% of patients are women, usually of reproductive age. Here is a case of a 35 year old woman who presented to us with a pedunculated, polypoidal mass arising from right labia majora since 6 years.

Keywords: Angiomyxoma, Mesenchymal tumor, Polypoidal mass

1. INTRODUCTION

Aggressive angiomyxoma is an uncommon and distinctive, mesenchymal tumor with a predilection for the pelvis and perineal regions in females and less frequently in males.[1] Steeper and Rosai first described its histologic characteristics and its tendency to local infiltration and recurrence in 1983. About 90% of patients are women, usually of reproductive age.[2] Here is a case of a 35 year old woman who presented to us with a pedunculated, polypoidal mass arising from right labia majora since 6 years.

2. CASE REPORT

A 35year old woman, para1 living 1 presented to us with a polypoid mass arising from right labium majora since 6 years, associated with pain since 15days. O/E

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she had a pedunculated skin covered polypoidal tissue measuring 8x7x2 cm with an ulcerated area of 6x6 cm with punched out edges and necrotic base. She underwent a wide local excision.

Histological examination of the specimen revealed a hypocellular tumor composed of spindle and stellate cells with an ill-defined cytoplasm, loosely scattered in a myxoid stroma without evidence of nuclear atypia and mitosis. Vessels of small to medium size with hyalinised wall noted.The tumor merged imperceptibly with overlying dermis. The stalk was free of tumor and showed thick walled vessels.

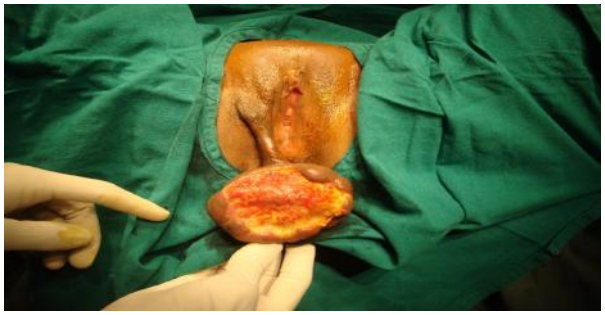

Fig 1: Polypoidal ulcerative mass from right labia majus

3. DISCUSSION

Aggressive angiomyxoma is an uncommon mesenchymal tumor. It characteristically grows slowly and insidiously and is regarded as an aggressive neoplasm because of its propensity to recur locally. The recurrence rate has been reported to be as high as 70% and most of these arise within two years. The age distribution is wide, with the peak incidence at 31 to 35years. Clinically the differential diagnosis includes a vulvar abscess/cyst, Bartholin abscess, Gartner's duct cyst, vaginal cyst/polyp, fibroepithelial polyp , lipoma, canal of Nuck hernia, pelvic floor hernia. It must be distinguished from benign proliferations with a low risk for recurrence, as well as from fully malignant myxoid tumors.

Microscopically it is a tumor with stellate and spindleshaped mesenchymal cells, embedded in a loose myxoid stroma with a few collagen fibers.There is absence of necrosis or mitotic figures. The cells are small and lack nuclear atypia.Small or medium-size veins and arteries within the tumor are often grouped together, some of them displaying medial hypertrophy. Blood vessels are often widely dilated,varying in caliber and usually larger than 10 mm in diameter and extravasation of red blood cells is common.[3, 4]

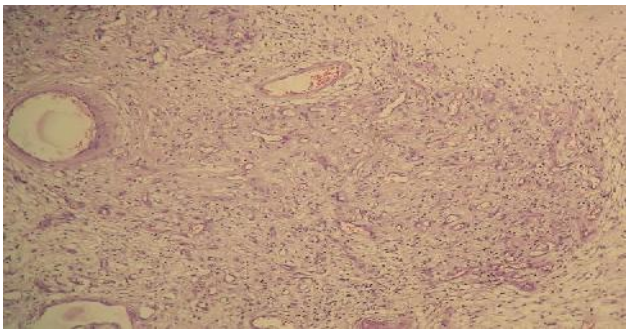

Fig 2: Microscopic appearance of Aggressive angiomyxoma

Immunohistochemically -It expresses different combinations of estrogen and progesterone receptors, vimentin, desmin, smooth-muscle actin, CD34, and CD44, but ***all are invariably negative for S-100, CEA, and keratin***.[5]

Genetics - Chromosomal translocation of the 12q13-15 band involving the HMGA2 gene has been described.

There is a wide range of differential diagnosis but the major ones are-

	AGGRESSIVE ANGIOMYXOMA	ANGIOMYOFIBROBLASTOMA	CELLULAR ANGIOFIBROMA
SIZE	usually >5cm	usually < 5 cm	small
BORDERS	infiltrative	usually well circumscribed	well circumscribed
VESSELS	numerous, medium to large sized, often hyalinized	Delicate capillary sized vessels	small to medium sized, often hyalinized
STROMA CELLS	Paucicellular spindle, stellate cells	Hyper and hypocellular spindle, plump, multinucleated cells perivascular accentuation	highly cellular spindle cells
STROMA	myxoid to collagenous	edematous to collagenous	wispy collagen bundles
BEHAVIOUR	recurrences in 30%	no recurrences	no recurrences

Recurrence - is notorious for local recurrence in approximately 70% of the cases after a period of 2 years postoperatively and has been reported 20 years postoperatively as well.

4. TREATMENT

The optimal treatment for AA is wide local excision with tumor free margin. Partial excision may be warranted when high operative morbidity is anticipated. Adjunct hormonal treatment with tamoxifen, raloxifene, and gonadotropin releasing hormone analogs (GnRHa) has been described with varying degrees of success. [3-5]

5. PROGNOSIS

Despite the morbidity associated with tumor recurrence and repeat surgeries, the prognosis for patients with AA is generally considered good. [6-8]

In spite of the benign nature of this neoplasm suggested by the histology, 2 cases of distant metastasis have been reported in the literature. This suggests that an aggressive angiomyxoma can no longer be considered a purely localized disease. It may act in a malignant fashion and produce benign metastases in a small percentage of cases.

6. REFERENCES

1. 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: 463–475
2. Smith HO, Worrell RV, Smith AY, et al. Aggressive angiomyxoma of female pelvic and perineum: review of the literature. Gynecol Oncol 1991; 43:79–85
3. Dahiya K, Jain S, Duhan N, Nanda S, Kundu P. Aggressive angiomyxoma of vulva and vagina: a series of three cases and review of literature. Archives of Gynecology and Obstetrics 2011; 283(5): 1145–1148.
4. Dierickx K. Deraedt W. Poppe, Verguts J, Aggressive angiomyxoma of the vulva: a case report and review of literature. Archives of Gynecology and Obstetrics 2008; 277(6): 483–487.
5. NX SunandW Li. Aggressive angiomyxomaof the vulva: case report and literature review. Journal of International Medical Research 2010; 38(4): 1547–1552.
6. Geng Junzu, MD, Cao Bofeng, MD, Wang Liping, MD. Aggressive Angiomyxoma: an Unusual Presentation, Korean J Radiol 2012;13(1):90-93
7. Metin Akbulut, Nese Ç. Demirkan, Nagihan Çolakoglu, Ender Düzcan. Aggressive angiomyxoma of the vulva: A case report and review of the literature. Aegean Pathology Journal 2006; 3: 1–4
8. Elkattah R, Sarkodie O, Otteno H, Fletcher A. Case Report Aggressive Angiomyxoma of the Vulva: A Précis for Primary Care providers. Case Reports in Obstetrics and Gynecology 2013; 1-4.