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CASE REPORT

Aggressive (deep) angiomyxoma of the vulva: Arare case report with review of literature

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ABSTRACT
Aggressive angiomyxoma is a rare, locally invasive mesenchymal stromal tumor which has a moderate-to-high risk for local relapse.
It arises in the soft tissues of the pelvis, perineum, andlower genital tract. It is unusual to metastasize. The most common affected group is of reproductive age. We present here a case of a 40-year-old female presenting with a large, fleshy, pedunculated mass on the vulva.
Key Words: Aggressive angiomyxoma, Vulval tumour, Mesenchymal tumor

INTRODUCTION

Steeper and Roasai originally reported aggressive angiomyxoma, a locally invasive mesenchymal stromal tumor, in 1983[1].
The underlying causes of AAM are still unknown at th is time. Aggressive angiomyxoma may be associated with chromosome alteration in 12q13-15 region[2,3].t(8;12) chromosomal translocation-induced aberrant expression of the DNAarchitectural f actor gene, HMGIC[4]. There have been less than 350 reported cases to date[5].
Estrogen and progesterone receptors are related to Angiomyxoma. So in pregnancy, it is likely to grow rapidly [6]. Occasional cases are seen in perimenopausal females and children. Here,we present a case of a 45-year-old female presenting with a large, fleshy, pedunculated mass on the vulva.

CASE REPORT

A 45-year-old woman presented to the gynecology department with a history of a mass on her left labium majora that had been steadily increasing in size for seven years. With The mass has superficial ulceration with serosanguinous discharge. She had no other menstrual complain.
On local examination, the left labium majora had a well-circumscribed, mass measuring 4 x 3 cm in size. The mass felt soft on the touch, nontender, pedunculated and nonreducible. The floor of the ulcer was covered in poor pale granulation tissue, and there was an overlaying 1x1cm ulcer. Ultrasonography (USG) of the local part revealed heterogeneous hyperechoic areas with peripheral vascularity and nonvascular central areas.With a clinical diagnosis of a vulvar fibroepithelial polyp or lipofibroma, she underwent local excision. Based on histology, sections showed stratified squamous epithelial lining, underneath stroma is hypocellular and edematous composed of small bland spindle cells with scant cytoplasm. Numerous blood vessels of varying size including thin walled capillary lining and thick walled arteries and extravasated red blood cells are also seen which were diagnostic of agressive angiomyxoma. (Figure1, 2)

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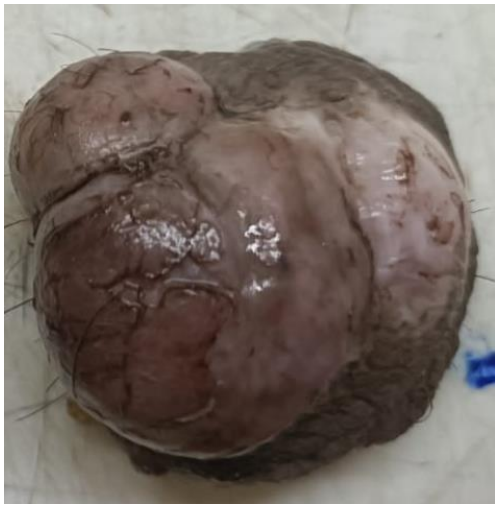

Fig 1: On a gross examination - soft, lobulated, and moderately well-circumscribed tumor with overlying skin.

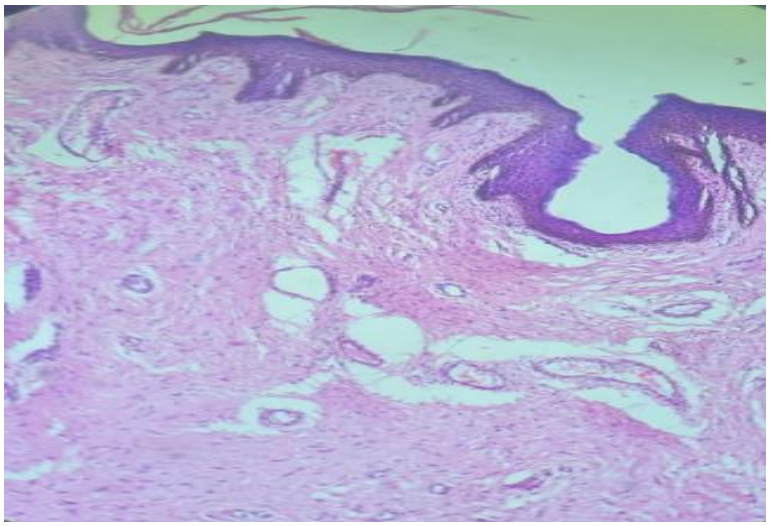

Fig 2: Stratified squamous epithelial lining, underneath stroma is hypocellular andoedematous− H&E ×100. H&E, haematoxylin and eosin.

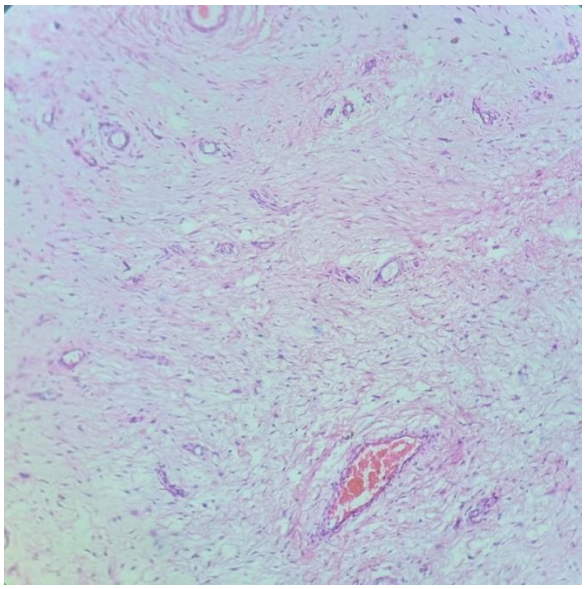

Fig 3: Thin and thick walled blood vessels with extravasated RBC's and edematous stroma.H&E ×400. H&E, hematoxylin and eosin.

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DISCUSSION

Superficial angiomyxoma are also known as cutaneous myxoma. Gross features are polypoid cutaneous mass arising from the superficial tissues. Common age group is predominantly in middle-aged male adults and may occur in the setting of the Carney complex. When it comes to females of reproductive age, the vaginal, perineal, and pelvic regions—especially the vulva—are considered aggressive, whereas the trunk, lower extremities, head, and neck are considered superficial. [7].

On a gross level, the skin-overlying tumors are soft, lobulated, and moderately well-circumscribed. The cutting surface of these tumors can be translucent, gelatinous, glistening, slimy, or colloid-like.[8]

Differential diagnosis includes angiomyoblastoma, myxoid neurofibroma, myxoma, spindle cell lipoma, myxoid liposarcoma, leiomyosarcoma, and botryoid rhabdomyosarcoma [9]

The radiological study of neoplasm is significant to determine the actual extension of the tumour; it is also useful for surgical planning and recurrence monitoring. Indeed, it is usually slow-growing and locally infiltrative, extending insidiously into adjacent soft tissues but rarely invading pelvic organs. The infiltrative behaviour and the absence of a well-defined capsule make complete removal difficult and contribute to the high rate of tumourrecurrence[10].

On H&E staining, the tumor cells are spindle or star-shaped. There is a mucus interstitial background. A large number of blood vessels are seen which are disordered and randomly scattered. Medium size blood vessels have thickened walls and do not appear to be in an anastomosis. The diameter and thickness of vascular tube range from fine arteries to larger arteries. [11].

CONCLUSION

Early diagnosis is essential for angiomyxoma due to it s aggressive nature and high risk for local recurrence.

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Conflictsofinterest
There are no conflicts of interest

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 Pathol1983;7:463-75.
2. Elkattah R, Sarkodie O, Otteno H, et al. Aggressive angiomyxoma of the vulva: a precis for primary care providers. Case Rep ObstetGynecol2013;2013:183725.
3. Srinivasan R, Mohapatra N, Malhotra S, et al. Aggressive angiomyxoma presenting as a vulval polyp. Indian J Cancer 2007;44:87–9.
4. NucciMR,Weremowicz S, Neskey DM, et al. Chromosomal translocation t(8;12) induces aberrant HMGIC expression in aggressive angiomyxoma of the vulva. Genes Chromosomes Cancer 2001;32:172–6.
5. Gaurav A, Gill P, Khoiwal K, Chowdhuri S, Kapoor D, Chaturvedi J. Aggressive angiomyxoma of the vulva—a rare entity: case report and review of literature. Int J Reprod Contracept Obstet Gynecol. 2020;9:2605. https:// doi.org/10.18203/2320-1770.ijrcog20202357.
6. Han-Geurts IJ, van Geel AN, van Doorn L, M den Bakker, Eggermont AM, Verhoef C. Aggressive angiomyxoma: Multimodality treatments can avoid mutilating surgery. Eur J Surg Oncol 2006;32:1217-21.
7. Zizi-Sermpetzoglou, A.; Myoteri, D.; Koulia, K.; Kontostolis, V.; Moschouris, H.; Dellaportas, D. Aggressive angiomyxoma of the vulva: A bizarre perineal lesion. Case Rep. Oncol. Med. 2015, 2015, 292304.
8. Lee CC, Chen YL, Liau JY, Chen CA, Cheng WF. Superficial angiomyxoma on the vulva of an adolescent. Taiwan J Obstet Gynecol. 2014 Mar 1;53(1):104-6.
9. Kondo, T. Aggressive angiomyxoma in the inguinal region: A case report. J. Med. Case Rep. 2010, 4, 396. [CrossRef]
10. Zamani, M.; Mollabashi, M.; Mehrabi, N.; Alizadeh, S. Aggressive angiomyxoma of vulva in 28-years old patient: A case report of second recurrence. Ann. Med. Surg. 2021, 69, 102706.
11. McCluggage WG. Recent developments in vulvovaginal pathology. Histopathology 2009;54:156–73.