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Case Report

A rare case of pedunculated vulval angiomyofibroblastoma

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ABSTRACT

A rare benign mesenchymal tumour that predominantly occurs in lower genital tract, more frequently vulvo vaginal areas of young to middle-aged women, rarely occurs in retroperitoneum. It needs to be differentiated from other benign conditions like Gartner's duct cyst, epithelial inclusion cyst, fibroepithelial polyps. It grows more slowly and is non tender, non invasive, and have least tendency for local recurrence. The histologic findings of the tumors are abundant thin-walled blood vessels with hypocellular and hypercellular areas. Almost all tumor cells have immunoreactivity for both desmin and vimentin. It also has estrogen and/or progesterone receptors, but staining for cytokeratin is negative. Here is a case of AMFB of the vulva occurring in a 62-year-old woman, involving the left labia majora. The patient described that her vulva mass grew over few years. The dimension of the tumor was measured as 18*12*4 cms. Excision biopsy done and sent for histopathology.

1. Introduction

Angiomyofibroblastoma is a benign mesenchymal tumour that occurs predominantly in subcutaneous tissue of vulvovaginal region of premenopausal women and very rarely in the inguinoscrotal region of men.[1] While majority of the tumors are encountered in the vulva, 10% to 15% of them occur in the vagina, occasionally in the perineum, inguinal region and retroperitoneum.[2] Differential diagnosis of mesenchymal neoplasms in the vulvovagina includes an array of lesions that are overlapping in both their morphology and immunohistochemistry like lipoma, epithelial inclusion cyst, fibroepithelial polyps and locally invasive aggressive angiomyxoma.[3]

2. Case Report

A 62-year old postmenopausal female, P_4L_4, presented with a painless vulval mass that started few years before, which had gradually grown to present size. She was a known hypertensive on medication. No significant past and family history.

Clinical examination showed a normal general physical examination and local examination showed a pedunculated mass, arising from lower part of left labia majora, soft to firm in consistency, measuring approximately 10*10 cms, pedicle measured approximately 8-10 cms.(as shown in Figure 1) Pelvic examination was normal.

2.1. Differential diagnosis

Vulval fibroma, vulval lipoma.

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2.2. USG

Solid heterogenous lesion from left labia measuring 18*10*4 cms with volume of 490 – 500cc with significant internal vascularity, probably angiomyofibroblastoma or vulval lipoma or labial sarcoma.

After relevant investigations, mass was excised and sent for histopathology.

2.3. Gross findings

Well demarcated, soft tissue mass measuring approximately 18*13 cms. Firm in consistency.

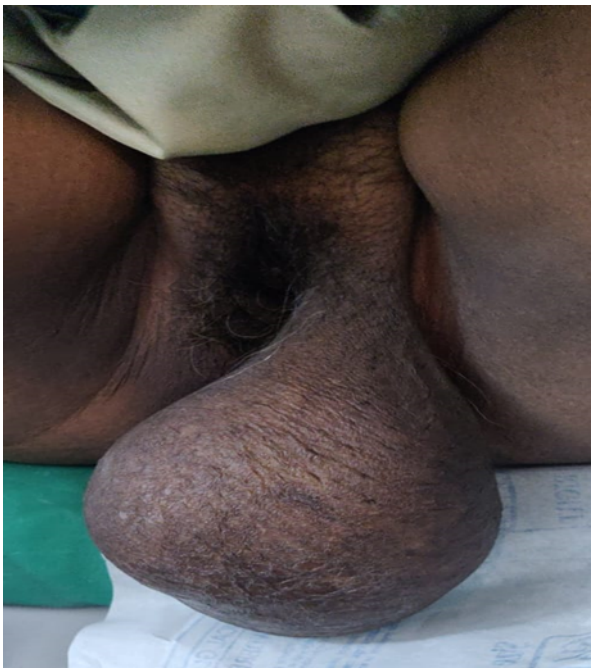

Fig. 1: Benign Angiomyofibroblastoma

2.4. Cut section

Bright yellow homogenous fatty tissue with fine fibrous capsule and trabeculae (as shown in Figure 2). Specimen sent for histopathology, reported as Angiomyofibroblastoma

2.5. HPE

Benign lesion with alternating hypercellular and hypocellular areas admixed with blood vessels (Figure 3 b). Spindle cells and plump stromal cells are seen aggregating around the vessels.(Figure 3 a) Bundles of muscle fibres and mature adipose tissue can be seen.(as shown in Figure 3 c)

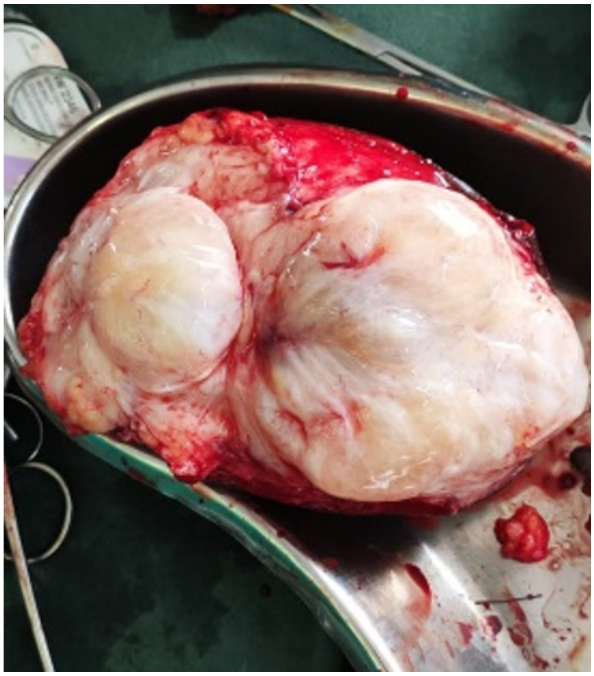

Fig. 2:

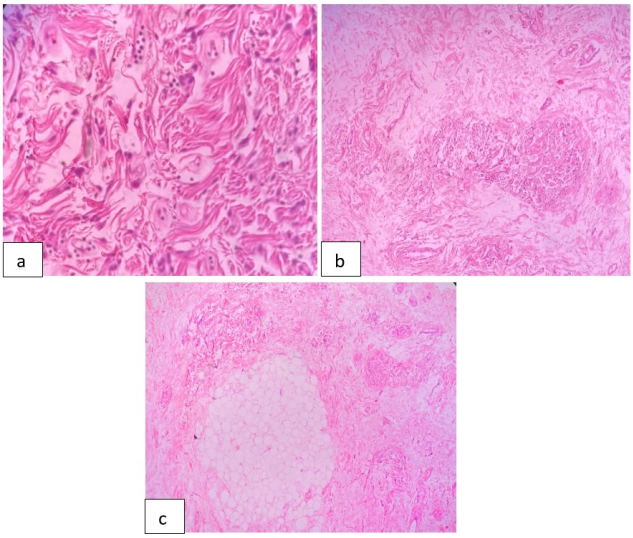

Fig. 3:

3. Discussion

1. AMF is a benign, soft-tissue tumor, its cause and pathogenesis are unknown at present. Since AMF is rare, no statistical data are available about its incidence in the general population. Most cases of AMF are benign, and only one case with sarcomatous transformation has ever been reported.[4]
2. On perineal USG, appear as a soft-tissue mass with inhomogeneous mixed echogenicity, which corresponds to the cellular inhomogeneity found on

histopathological examination.

3. Immunohistochemistry-tumors express estrogen and progesterone receptors, which suggests that it might have originated as a neoplastic proliferation of hormonally responsive mesenchymal cells, also positive to desmin, vimentin and smooth muscle actin.[5,6]

4. Conclusion

Although the exact nosologic position of angiomyofibroblastoma is still surrounded by some controversy and requires further elucidation. Simple excision appears sufficient in its management. Whereas wide excision after histological examination is needed for the management of the aggressive angiomyofibroma which is associated with a high risk of local recurrence and infiltration.

5. Source of Funding

None.

6. Conflict of Interest

None.

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