

Multiple Angiofibromas of Vulva in post menopause – A Case Report.

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INTRODUCTION:

Cellular angiofibroma was first described by Nucci et al. in 1997. Chelins in 1847, first recorded the lesion and Friedberg in 1940, coined the term “angiofibroma”. Cellular angiofibroma (CAF) is a rare benign mesenchymal lesion usually found in the vulvovaginal region. It is composed of vessels and fibrous connective tissue. . Unlike most of the other site specific genitourinary mesenchymal lesions, cellular angiofibroma has a marked predilection for the vulva especially labia majora with only occasional examples reported in the vagina .

Clinical presentation:

A 52 years old postmenopausal nulliparous lady presented in the outpatient department with multiple painful swellings, on the vulval region which was gradually increasing in size for the last 12 months. **B**iopsy from the mass was taken at a peripheral centre and reported as angiofibroma in histopathology.

General and abdominal examinations normal except for pallor.

On local examination, multiple polypoidal masses arising from her mons pubis extending to the left labia majora with some areas of surface necrosis and discoloration. They were tender and felt firm in consistency. On vaginal examination uterus atrophic, mobile, fornixes were free. The histopathology was reviewed in our Onco-pathology department and diagnosis confirmed. Routine blood and imaging were normal.

Management:

Local excision with flap repair was planned along with plastic surgery department during tumor board.

The total lesion was excised from the mons pubis and the left labia majora. The plane of dissection was along the camper’s fascia. The defect was large enough to need bilateral lotus petal flap and additional split thickness skin grafting (from the left thigh). On grossing, the excised specimen was of 15x9x4cm dimensions. There were 4 solid nodules arising out of the outer surface, largest measuring7x4.5x3.5cm. Cut section showed solid lesions with uniform fleshy consistency. Post-operative period was uneventful. Final histopathology report was consistent with angiofibroma of vulva. Immunohistochemically it was CD 34 positive and S100 negative.

Vulvovaginal tumors are uncommon and it includes a spectrum of fibroepithelial tumors, soft tissue specific and non-specific tumors. CAF, myofibroblastoma and angiomyoibroblastoma are usually considered as specific soft tissue tumors of the vulvovaginal area. These tumors may show overlapping morphological, immunohistochemical and cytogenetic features, and thus it is important to differentiate them. CAF occurs approximately 10 years later in life than myofibroblastoma or angiomyoibroblastoma or aggressive angiomyxoma , mostly observed in the fifth decade . They are usually small, well circumscribed, asymptomatic stromal tumors and occur in both the genders with equal preference . Though CAF are slow growing, they can reach enormous sizes before they present. The usual differential diagnoses include, Bartholin’s cyst in 48% of cases, a non-specific solid mass (28 %), vulvar cyst (12%), fibroid (8 %) or lipoma (4 %) .

Etiopathogenesis is indefinite with some authors suggesting that these tumors are stem cell derived with a capacity for adipose or myofibroblastic differentiation under the influence of microenvironments, hormones, cytokines and growth factors. CAF is composed of two principal components: bland spindle cells and prominent small to medium-sized vessels with mural hyalinization Immunohistochemically tumor cells are vimentin positive and almost 60 % cases are CD 34 positive.. Typically, they don’t express S-100 protein although a discrete staining for actin, desmin, or EMA have been reported.



Relevance:

It is imperative to keep these unusual cases in consideration and distinguish them from malignancy in order to avoid overtreatment

HP: High Power view showing proliferation of bland looking spindle cells in short fascicles and haphazard manner

