

Cellular Angiofibroma of Vulva: A Rare Case Report

Amit Basu¹, Rajni Bansal², Upasna Sharma³, Srabani Auddy Basu⁴

ABSTRACT

Aim: To describe clinical, histological, and immunohistochemical features of cellular angiofibroma, a rare mesenchymal tumor of vulva.

Background: Cellular angiofibroma are rare mesenchymal tumors. These tumors have two principal components spindle cells and hyalinized stroma with small to medium sized vessels. Cellular angiofibroma (CAF) are benign in nature. Cellular angiofibroma commonly occurs after fifth decade. Characterization requires histological and immunohistochemical evaluation.

Case description: We report a case of 65-year-old post-menopausal woman presenting with a painless and gradually enlarging mass in clitoral region of vulva near external urethral meatus. A simple excision of mass was performed. Gross examination of the mass showed a well circumscribed lesion of 5.5 cm × 5.5 cm. Histological and immunohistochemical analysis established a diagnosis of cellular angiofibroma. The tumor can recur but distant metastasis of the tumor has not yet been described in the literature.

Conclusion: Cellular angiofibroma is a rare benign tumor of vulva for which simple excision is adequate treatment. Differentiation of this tumor from other sarcomatous lesions can be done with histological and immunological evaluation.

Keywords: Cellular angiofibroma, Immunohistochemistry, Histopathology, Mesenchymal tumors, Vulva.

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INTRODUCTION

Soft-tissue sarcomas of the vulva are rare class of gynecologic malignances. Of these, cellular angiofibroma is a rare benign mesenchymal lesion with a predilection for the genitourinary region. They were first described in 1997.¹ Cellular angiofibroma is characterized by a spindle cell component and abundant small- to medium-sized thick-walled vessels.²

CASE REPORT

A 65-year-old post-menopausal female presented to our OPD with a history of a non-tender vulvar mass involving clitoris which was enlarging in size and first noticed 3–4 months back. She had no complaint of pain, any discharge from the mass, any difficulty in urination, burning sensation in urine, any recent weight loss. The patient gave a history of similar lesion excised from the same location 20 years back for which no record was available with her.

On physical examination, a 5.5 cm × 5.5 cm well circumscribed, firm, non-tender mass at 12 o'clock position was found on labia which involved the clitoris, very near to the external urethral meatus and was mobile over underlying bone. On palpation, no inguinal lymphadenopathy was appreciated. The overlying skin was normal in appearance.

The patient underwent a wide local excision of mass with adequate margins. It was a firm to soft solid mass involving the clitoris till 0.5 cm from the external urethral meatus with no fixity to the deeper bones. The lesion was arising from the corpora of clitoris (Fig. 1).

Patient was discharged on the fifth post-operative day in a stable condition.

Differential diagnosis of this condition includes vulvar inclusion cysts, Bartholin's gland cyst, submucosal cyst, vulvar soft tissue neoplasms, and vulvar cancer.

Gross histopathological findings showed a partially cut open well circumscribed ovoid mass of size 5.5 cm in its greatest axis. The cut surface was grayish.

^{1,2}Department of Obstetrics and Gynaecology, BR Singh Railways Hospital, Sealdah, Kolkata, West Bengal, India

³Department of Pathology, Government Medical College, Patiala, Punjab, India

⁴Department of Anaesthesiology, BR Singh Railways Hospital, Sealdah, Kolkata, West Bengal, India

Corresponding Author: Rajni Bansal, Department of Obstetrics and Gynaecology, BR Singh Railways Hospital, Sealdah, Kolkata, West Bengal, India; Phone: +91 8239330092, e-mail: rbrajnibansal@gmail.com

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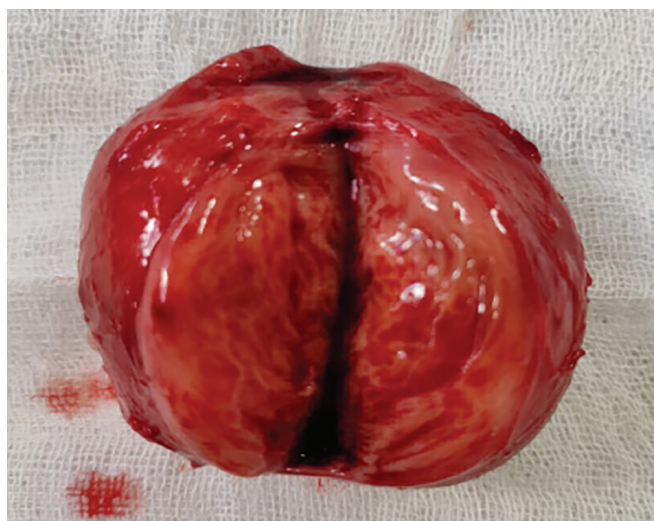


Fig. 1: Cut section of the excised lesion

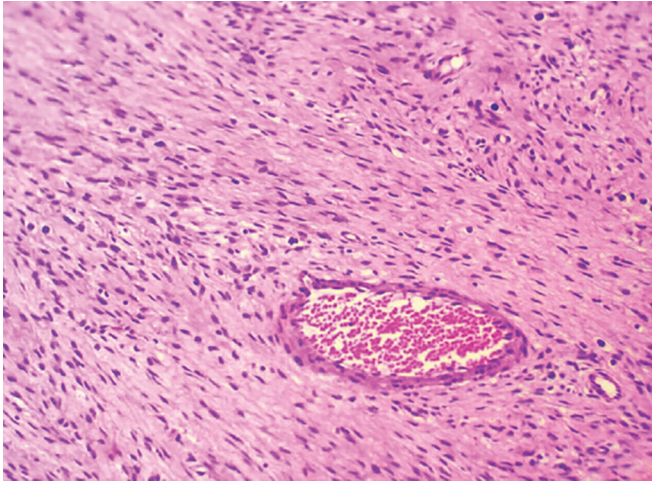


Fig. 2: Histopathological appearance of tumor

Microscopic examination revealed a moderately cellular spindle cell neoplasm composed of haphazardly arranged oval to spindle cells with bland vesicular nuclei and pale eosinophilic cytoplasm. The stroma was collagenous. Mitosis was sparse (2–3/10 hpf). Dilated capillary sized blood vessels noted. Mast cells were present in the stroma. The tumor abuts the circumferential resection margins at places. These findings gave an impression of unclassified spindle cell neoplasm (Fig. 2).

Immunohistochemical analysis showed a cellular spindle cell neoplasm composed of uniform, plump spindle-shaped cells in a fibro-collagenous stroma with interspersed small sized blood vessels with ecstasic lumina. The lesional cells had plump to ovoid nuclei with inconspicuous nucleoli and pale eosinophilic cytoplasm. Mitotic figures were not discernible. Mast cells were prominently seen scattered in the neoplasm. Lesional cells expressed ER and were immunonegative for desmin, SMA, S-100 PROTEIN, and CD34. The tumor cells showed loss of nuclear RB-1 protein expression. Tumor reached up to inked margin of the excision. The final impression from the above findings was cellular angiofibroma.

DISCUSSION

Tumors primarily arising from the vulvovaginal area are relatively rare and they include soft tissue specific and non-specific tumors, as well as a spectrum of fibroepithelial tumors.^{3,4} Cellular angiofibroma are rare benign stromal tumors, affects both genders equally and mostly found in vulvovaginal region. Only few reports of these lesions and that too in form of single patient case reports are available in literature.

Histologically, CAF predominantly are based in subcutis; rarely in dermis. These are usually well circumscribed; rarely infiltrative. Fibrous pseudo capsule is present in a subset.

These are composed of two principal components. Tumor cells which are small, monotonous spindle cells with bland, ovoid to fusiform nuclei, and are arranged in short intersecting fascicles.

Bland multinucleated cells are common finding. Mitoses is typically a rare finding (<1 per 10 hpf) but occasionally brisk mitosis can also be seen (>10 per 10 hpf). Rare cases show focal or diffuse atypia or discrete areas of sarcomatous transformation. Tumor stroma and vasculature stroma can be myxoid, edematous, fibrous, or hyalinized. Usually, CAF has abundant medium sized, thick walled, and hyalinized vessels. Stroma is arranged in short wispy collagen bundles. Minor adipocytic component can be seen in around ~50%; rare cases show prominent adipocytic differentiation and when present; it mostly occurs in the periphery of lesion. The mast cells may be conspicuous. Necrosis and hemorrhage are characteristically absent.

Immunohistochemically, CAF are vimentin positive; CD34 positive in 60% cases.⁵ Most cases are usually estrogen (ER) and progesterone receptor (PR) positive. Characteristically, they do not express S-100 PROTEIN, actin, desmin, or EMA, although a discrete staining for the last three markers has been reported.⁵

Molecular/cytogenetically, these tumors have monoallelic loss of RB1 and FOXO1A1 genes which is seen due to the deletion of the 13q14 region.

Cellular angiofibroma has characteristics of a benign tumor and simple local excision with clear margins is sufficient and is the treatment of choice. Local recurrence is very rare even if positive margins. So-called sarcomatous transformation does not increase the risk of recurrence. No distant metastasis case has been reported.

CONCLUSION

Vulvovaginal sarcomas, particularly, spindle cell sarcomas are rare, and clinical diagnosis and distinguishing from other neoplasms is difficult. Surgical excision followed by histopathologic and immunohistochemical evaluation is necessary for diagnosing and distinguishing them from other mesenchymal lesions of the vulva.

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