

Cellular Angiofibroma: A Rare Vulvar Neoplasm Distinct From Aggressive Angiomyxoma

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ABSTRACT

Cellular angiofibroma (CA) is a rare benign mesenchymal neoplasm of the genital region of both the genders. In women, it arises in late reproductive age and can be cured by complete local excision. It is characterized by a bland spindle cell population and numerous small- to medium-sized vessels with hyalinization. We report a rare case in a postmenopausal woman with a view to highlight the importance of differentiating it from similar more aggressive tumours like aggressive angiomyxoma. A 56 year old woman presented with superficial painless swelling of the vulva and the mass was surgically excised. Histological examination revealed spindle cell proliferation and hyalinised blood vessels in a loosely cellular stroma. Immunohistochemistry revealed positivity for vimentin and CD34 and positivity for ER & PR.

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Introduction

Cellular Angiofibroma (CA) is a rare benign mesenchymal neoplasm of the genital region of both the genders. It arises in women of late reproductive age ^[1, 2] and can be cured by complete local excision. It is characterized by a bland spindle cell population and numerous small- to medium-sized vessels with hyalinization. These tumours can be mistaken for more aggressive tumours and one should be aware of similar tumours presenting as vulvar swellings and overlapping histological features. We report a case in a postmenopausal woman with a view to highlight the importance of differentiating it from similar more aggressive tumours.

Case Report

A 56 year old lady presented with a painless vulvar swelling of one year duration. Her general and systemic examination was normal. Routine hematological and biochemical investigations were within normal limits. Per operatively a well-defined mass was seen on the vulva and was excised. Postoperative period was uneventful and there was no recurrence.

Gross Examination: Gross examination of the tumour revealed a well circumscribed tumour measuring 7.4x4.5x2.3cm, with a gray surface. (Fig1). Cut section of the tumour also revealed a well circumscribed grey white tumour. There were no areas of haemorrhage or necrosis.

Microscopic Examination: Histological examination showed a well circumscribed tumour composed of uniform spindle cells arranged in short fascicles. Cells had indistinct cytoplasm and blunt to wavy nuclei. Many small to medium sized vessels with thick walls were interspersed in these spindle cells. Thin collagen bundles were seen along with mild lymphocytic inflammatory infiltrate. There were no mitotic figures. No necrosis or atypia was noted (Fig2, 3).



Fig. 1: Gross photograph showing well circumscribed gray white tumour with prominent blood vessels on surface.

Immunohistochemistry: The tumour cells were positive for vimentin and CD34, ER and PR. (Fig 4, 5, 6, 7) The tumour was negative for S-100 protein, actin, desmin and EMA. Histomorphology and immunohistochemistry was compatible with cellular angiofibroma of the vulva.

Discussion

Cellular angiofibromas are rare tumours that present clinically as subcutaneous vulvar swellings in women of reproductive age group. CAs were first described by Nucci et al in 1997 as tumors with a histomorphology consisting of bland spindle cells along with small to medium hyalinized vessels. Mature adipose tissue may be seen in the tumours. ^[3]

These tumours are site specific and one should be able to differentiate these from other spindle cell lesions in the perineum. ^[4] Aggressive tumours like aggressive angiomyxoma may present like CA. Site specific tumours like angiomyofibroblastoma, spindle cell lipoma, leiomyoma, solitary fibrous tumor and perineurioma may also mimic CA.

Cellular Angiofibromas are vimentin positive. CD34 positivity is seen in 60% of the cases. Smooth muscle actin and desmin positivity is less with 20% and 8% of the cases showing positivity. ^[5]

Aggressive angiomyxoma is an infiltrative tumour with spindle cells in a myxoid stroma. This is larger than cellular angiofibroma. Vessels are medium-sized to large and show hyalinization. "Myoid bundles" composed of collections of smooth muscle are seen around blood vessels. Features such as an infiltrative margin and myxoid stroma helps in differentiating this from CA. Immunohistochemically these tumours are positive for desmin and actin in contrast to cellular angiofibroma and negative for CD34. ^[5,6]

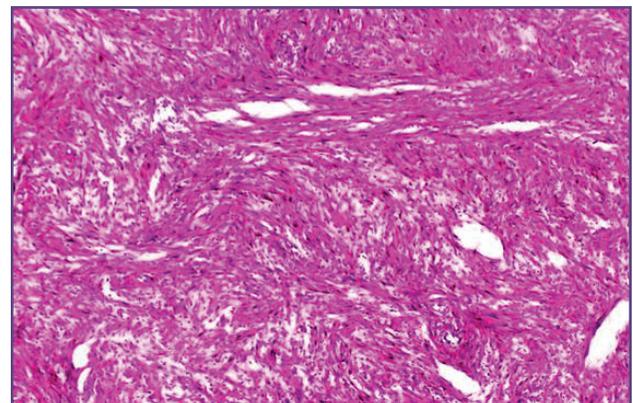


Fig. 2: Microphotograph showing short fascicles of spindle cells with hyalinized blood vessels. (H & E, X100)

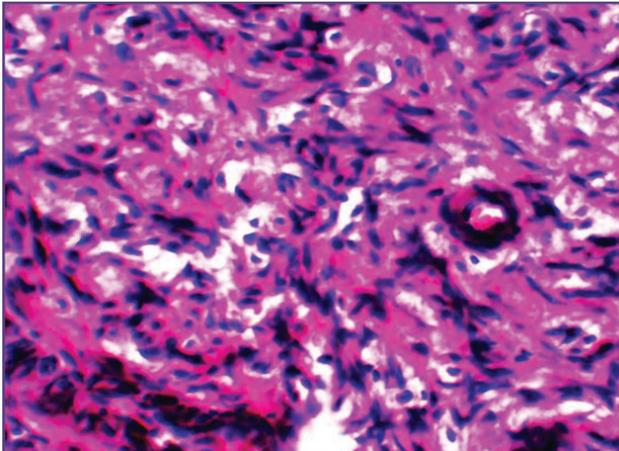


Fig. 3: Microphotograph showing spindle cells with eosinophilic cytoplasm and blunt to wavy nuclei and hyalinized blood vessels. (H & E, X400)

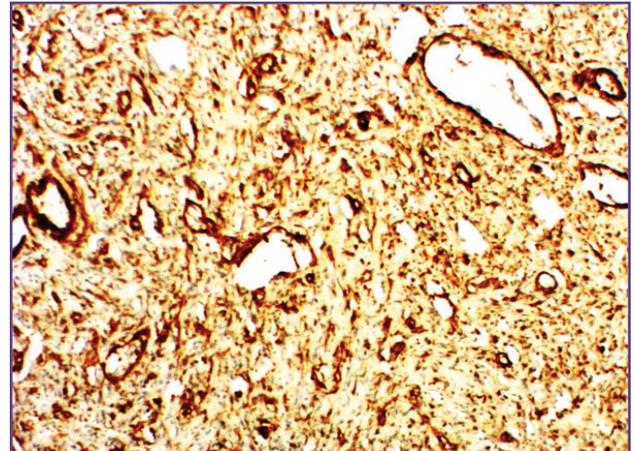


Fig. 4: Microphotograph with CD34 Positivity in both the endothelial cells and spindle cells.

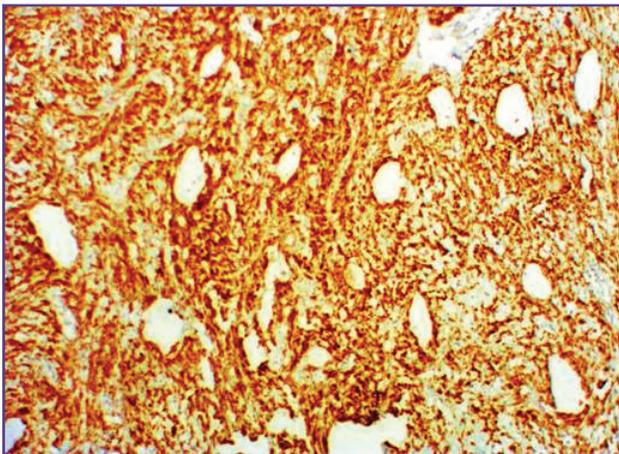


Fig. 5: Spindle cells and vessels showing positivity for vimentin.

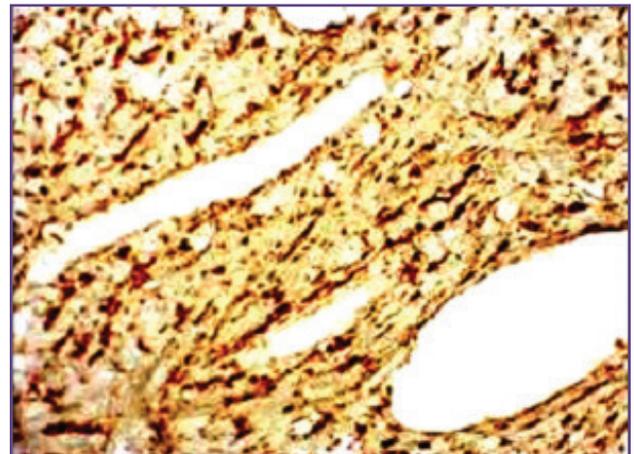


Fig. 6: Immunopositivity for ER in spindle cells.

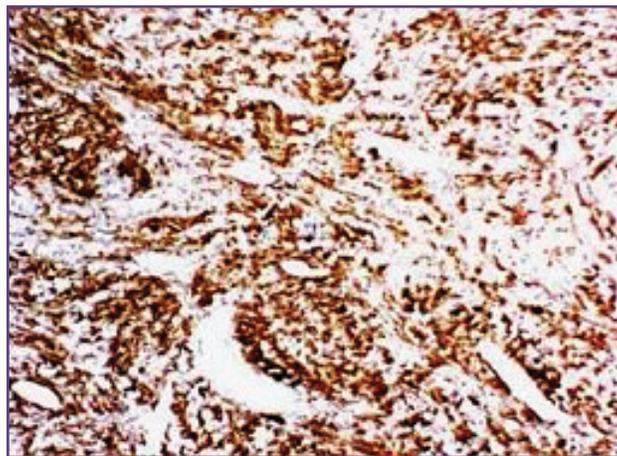


Fig. 7: Spindle cells showing positivity for PR.

Another tumour, which presents in the same site and with similar clinical features is angiofibroma (AMF). This is a benign, predominantly superficial circumscribed tumor. It can be differentiated from CA by the presence of alternate areas of cellularity. In addition to spindle cells, it also has epithelioid cells. Vessels in the tumour are capillary sized. The tumour expresses vimentin and desmin and occasionally smooth muscle actin and CD34.

Spindle cell lipoma is a lobulated, well circumscribed tumour showing admixture of spindle cells in fascicles and mature adipocytes. Spindle cells have poorly defined cytoplasm with uniform nuclei.^[7]

Solitary fibrous tumor is positive for CD34, CD99 and bcl-2 positive, but generally negative for S-100, actin, desmin.

Dermatofibrosarcoma protuberans (DFSP) is an uncommon slow growing fibroblastic tumour of the dermis. It most commonly occurs on the trunk but may occur in the vulva. It occurs in women in fourth or fifth decade and has a tendency for local recurrence. Histological features which help in differentiating this tumour from CA are poor circumscription and infiltration into subcutaneous adipose tissue in a lacy pattern. Histological examination shows irregular short intersecting bands of spindle cells forming a storiform pattern. It is separated from overlying epidermis by a grenz zone, and entraps adnexal structures.^[8] This tumour is positive for CD34 and negative for factor XIIIa and S100.

Perineurioma is a well circumscribed subcutaneous mass. It is characterized by variable cellularity and strikingly whorled, or storiform growth patterns. Cells are positive for EMA. Around 50% cells are CD34 positive and some cases may be S100 positive.

Leiomyomas of the vulva are clinically similar to cellular angiofibroma as they generally occur in the reproductive age group. Histomorphology is also similar, characterized by spindle cells in short fascicles. These tumours are positive for Smooth muscle actin.

Some studies have suggested a link between cellular angiofibroma, spindle cell lipoma, and mammary-type myofibroblastoma i.e. all the three entities are spectra of one entity depending upon the location of the tumour.^[9]

Diagnostic difficulty also arises sometimes due to variation in histomorphology of CA as described in a few studies. Some studies have documented increased mitotic figures in a few of these tumours.^[10]

Atypia has also been described. One of the studies have also documented sarcomatous areas in the tumour^[5]

In another study features like presence of adipose tissue, stromal mast cells, stromal lymphoid aggregates, scattered multinucleate cells, hypocellular hyalinized areas, myxoid areas and focal areas of marked cellular atypia reminiscent of symplastic change within a uterine leiomyoma have been described.^[10] In these cases immunohistochemistry helps in arriving at a diagnosis. We did not find any of these features in our case.

Conclusion

To conclude, cellular angiofibroma is a rare benign tumour and should be considered in the differential diagnosis of vulvar soft tissue tumors. Characteristic morphological features supplemented by immunohistochemistry helps in arriving at a diagnosis and differentiating from aggressive tumours like aggressive angiofibroma. Immunohistochemically, these cases are characterized by vimentin positivity and negative staining with smooth muscle markers which assist in excluding many of the other vulvovaginal mesenchymal lesions which enter into the differential diagnosis.

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Competing Interests

None declared

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