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Case Report

# Cellular Angiofibroma a Rare Tumor: A Case Report from Oman

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**Received:** November 12, 2018; **Published:** January 31, 2019

## Abstract

**Background:** Cellular angiofibroma (CA) is a rare benign mesenchymal neoplasm. It was first described in 1997. Since then and up to 2014, only 79 cases have been reported worldwide. Cellular angiofibroma incidence is equal in both genders and but presents in middle-aged women but older men. Vulvovaginal and inguinoscrotal regions in females and males respectively are the commonest sites of cellular angiofibromas.

**Case Description:** We report a 39 year old lady who presented to our institution with a painless left labial mass that has been gradually increasing in size for the past 6 months. Examination revealed a non-reducible, mobile, fluctuant 6 x 10 cm left labial mass with no signs of local inflammation. Imaging was suggestive of an aggressive angiomyxoma. She underwent simple excision and histopathology diagnosis was consistent with cellular angiofibroma. There is no evidence of local recurrence at 6 months follow up.

**Conclusion:** Cellular angiofibroma usually has a benign and excellent prognosis. It is important to differentiate from other aggressive soft tissue tumors with similar presentation. Histopathological examination after simple excision, with the aid of immunohistochemistry will achieve an accurate diagnosis. Data for long-term follow up are limited and the need for such data is important to ensure a totally benign course of the tumor. A few cases of atypia, sarcomatous transformation and recurrence are reported in the literature.

***Keywords:*** *Cellular Angiofibroma; Soft Tissue Tumor; Aggressive Angiomyxoma; Vulva*

## Background

Cellular angiofibroma (CA) is a rare, benign mesenchymal neoplasm. It was first described by Nucci., *et al.* in 1997 [1]. Cellular angiofibroma arises equally in both genders [2-4] and usually arises in the inguinoscrotal or vulvovaginal regions [5,6]. It is difficult to differentiate CA from other vulvar soft tissue tumors on clinical examination or imaging. The diagnosis is only established on histopathological examination. Local excision is the treatment of choice. We report a case of cellular angiofibroma with initial imaging that was suggestive of an aggressive angiomyxoma and illustrates the importance of histopathological examination to differentiate between aggressive and benign vulvar masses.

## Case Description

A 39 years old lady was referred from the Gynaecology to the General Surgery clinic with a painless left labial mass that has been present for the past 6 months. There was no associated increase in size, fever, discharge, loss of appetite or weight loss. There was also no urinary or bowel symptoms. Neither the patient nor her family had a history of breast or gynecological cancer. She had been married for 15 years and had not been on hormonal therapy. Examination revealed a non-reducible left labial mass measuring 6 x 10 cm which was freely mobile and fluctuant with no signs of inflammation. There were no palpable inguinal lymph nodes bilaterally. Systemic examination was otherwise unremarkable.

An ultrasound exam performed in the Gynaecology clinic showed a 6 x 7 cm left labial mass underneath the skin. A subsequent CT scan of the abdomen and pelvis showed a left labial subcutaneous enhancing mass suggestive of an aggressive angiomyxoma or neurofibroma. An MRI scan was next performed (Figure 1) and showed features suggestive of an aggressive angiomyxoma.

The patient underwent a simple excision with complete removal of the labial mass. Macroscopically, the lesion was well-defined and measured 11.5 cm in the greatest diameter with a solid tan cut surface. Microscopy showed a cellular, sharply demarcated neoplasm composing of short interlacing fascicles of spindles cells separated by short collagen fibers. Numerous small and medium-sized blood vessels were noted throughout the neoplasm. The spindle cells had bland appearing nuclei and eosinophilic cytoplasm. Occasional atypical and multinucleated cells were seen. There was no evidence of mitosis or tumor necrosis. On immunohistochemistry, the neoplastic cells were diffusely positive for CD34 and negative for SMA, Desmin, and ER. The morphology and immunoprofile was consistent with Cellular Angiofibroma. She did well postoperatively and is current on follow-up with no signs of recurrence after 6 months.

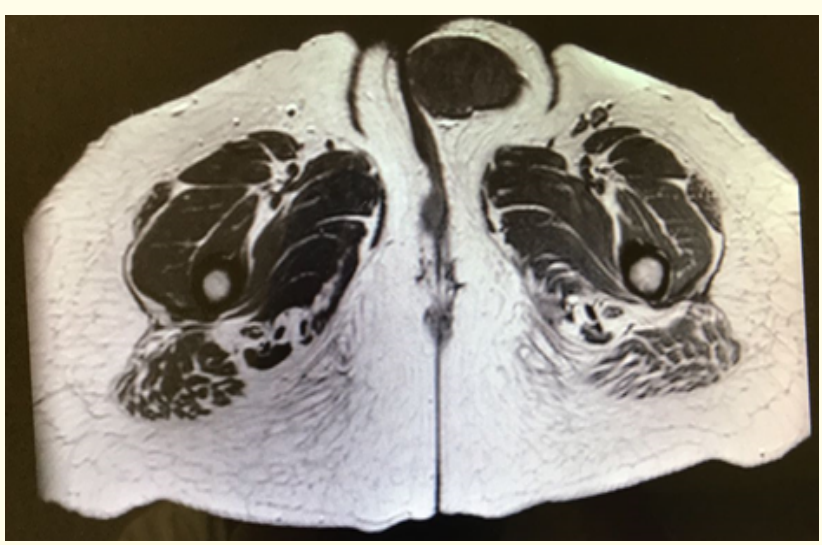

***Figure 1:*** *MRI showed a soft tissue mass in the left labia.*

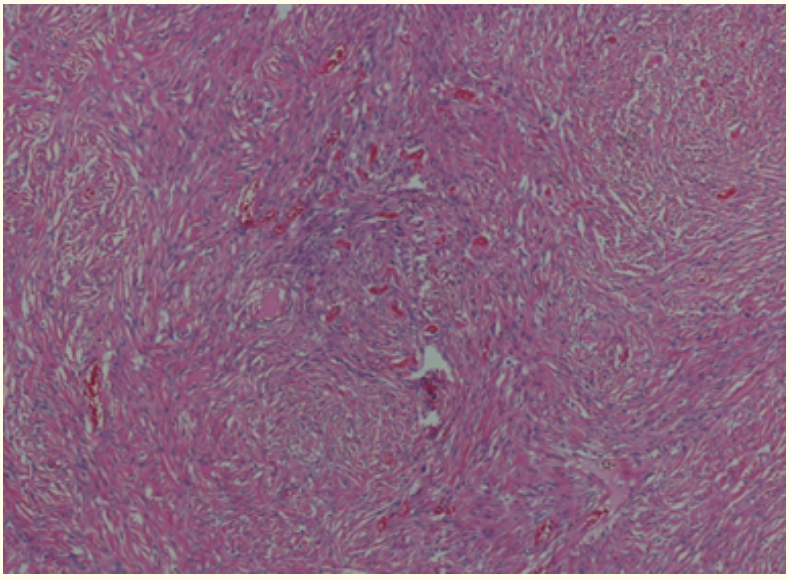

***Figure 2:*** *Short fascicles of spindle cells with multiple small and medium-sized blood vessels (H&E).*

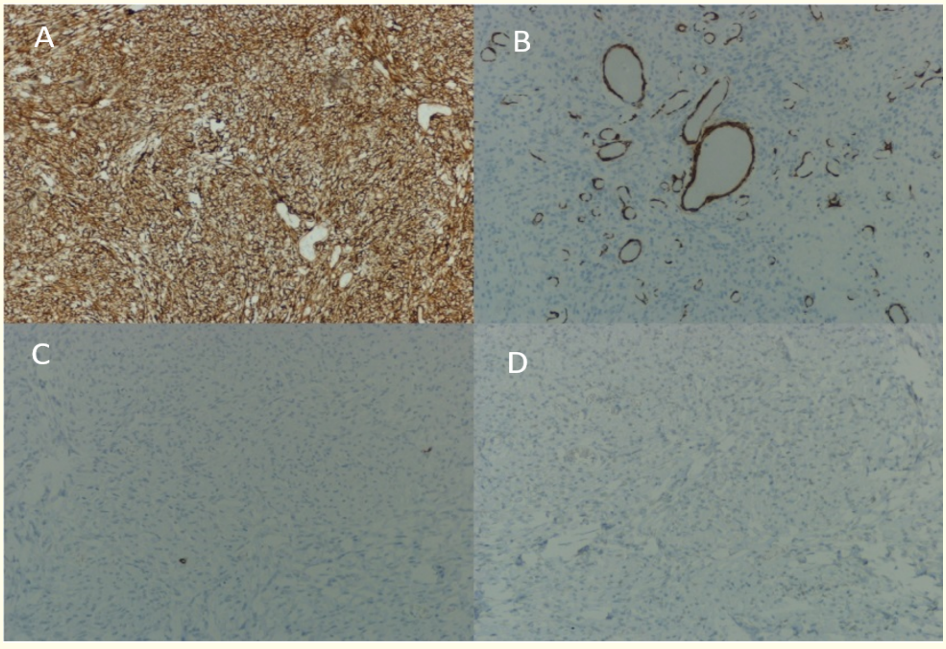


***Figure 3:*** *Immunohistochemistry: a) CD34, b) SMA, c) Desmin and d) ER. (IHC).*

## Discussion

Cellular angiofibroma (CA) was first described by Nucci., *et al.* in 1997. They reported four cases with distinctive features and came up with the term 'Cellular angiofibroma' [1]. CA arises equally in both genders [2-4] and usually develops in the inguinoscrotal or vulvovaginal regions. Extra-genital locations have also been reported in the literature, and these include regions such as the chest, abdomen, breast, retroperitoneum, knee, eyelid, anus and oral mucosa [4-7]. In females, CA commonly presents in the fifth decade of life with a painless, asymptomatic mass that gradually increases in size over an average of 1 - 2 years [8,9]. The size ranges from 0.6 - 12.3 cm [6], though a later case report discussed about a 20 cm sized CA [3]. In one case report, the patient had an atypical presentation with a painful mass [9].

Differential diagnosis include an inguinal hernia, Bartholin's cyst, vulvar cyst, leiomyoma or lipoma [6]. It is important to exclude an inguinal hernia as the operative approach is quite different. Current imaging techniques cannot distinguish a CA from other soft tissue tumors. Histopathological examination is needed to confirm the diagnosis, with immunohistochemistry playing a vital role [9].

Gross pathological examination of CAs usually reveals a white or yellowish nodular and mostly solid mass [6,8]. Macroscopically, it is usually found as a well circumscribed mass in the superficial soft tissue, though extension to surrounding tissues can also be seen [6]. There is a single reported case of a hemorrhagic foci in the CA, with no reports of necrotic foci thus far [6]. Microscopically, CA has two distinctive features, namely that of bland spindle cells and prominent small-medium sized vessels, some with hyalinization of the vessel wall. The stroma can be oedematous and may contain variable amounts of collagen fibres and inflammatory cells. In some case reports, hypocellular areas or scant mature adipocytes can be present [2,5-7]. CA can show a variable range of mitotic activity [6]. S-100 is negative in all cases [5,6], CD 34 is positive in approximately 45 - 60% cases [6,8,10]. Desmin, vimentin, ER, and PR are positive in some cases [5,6,10].

The differential diagnosis of CA include other mesenchymal neoplasms such as aggressive angiomyxoma (AA), angiomyofibroblastoma (AMF), myofibroblastoma, spindle cell lipoma, smooth muscle tumors and perineuroma. The spindle cell lipoma has same the bland spindle cell findings as in CA, though the latter has additional numerous blood vessels with thickened, hyalinized wall [5,7]. AAs are usually deep-seated tumours and tend to be locally destructive and display local recurrence. Both CA and AA show no metastasis risk [2,5,6]. Microscopically, AAs are characterized by small clusters of smooth muscle cells surrounding or 'spinning off' from blood vessels [7]. AAs can be positive for desmin and actin [5]. AMFs show similarity to AFs in that both are well circumscribed and well demarcated tumors, though the former is characterized microscopically by the presence of multinucleate cells and epithelioid or plasmacytoid cells which tend to aggregate around vessels [2,6]. Smooth muscle tumors can be distinguished from CAs by morphology and immunohistochemistry as they express muscle markers. Myofibroblastoma, spindle cell lipoma and CAs are probably a spectrum of one entity, as they share the same cytogenetic aberration (del of 13q14.2 region) as confirmed by FISH analysis [2,6,8].

Simple excision of the mass with clear margins is the treatment of choice for CA. En-bloc resection of the mass with its accompanying capsule ensures both hemostasis, with CAs being highly vascular neoplasms, and prevention of local recurrence. Although there is limited long-term follow-up data, there is one reported case of local recurrence 6 months after simple excision with clear margins [11]. No metastasis have been directly attributed to CA [6,12], though there is a single case report of a patient who passed away 27 months after diagnosis from metastatic disease of unknown origin [6]. Sarcomatous transformation and atypia have been reported in CAs, though there remains no evidence to support a tendency for recurrence or distant metastasis [7,13]. In these cases, p16 tumor suppressor gene overexpression was also noted, unlike the usual cases of CA [6]. Despite so, the clinical course of CA is generally benign with excellent prognosis [5,8,9].

## Conclusion

Mesenchymal lesions of the vulva and perineum can be benign or malignant. The cellular angiofibroma is a benign neoplasm that has a good prognosis. Exclusion of other potential neoplasms like aggressive angiomyxoma is crucial. Histopathology review is needed to confirm the diagnosis. The pathogenesis of the cellular angiofibroma is still not fully determined and current hormonal and immunotherapy suppression regimes will need further evaluation.

## Conflicts of Interest

We have no conflicts of interest to disclose.

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**Volume 6 Issue 2 February 2019**

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