Case Report

Vaginal Cellular Angiofibroma: Report of a Rare Case with Literature Review

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Abstract

Cellular angiofibroma (CA) is a relatively recently described, rare, benign soft tissue tumor that predominantly occurs in the vulvoperineal region. It was first described in 1997 as a mesenchymal-origin tumor composed of connective (fibrous) tissue and blood vessels. We report a case of vaginal CA in a 40-year-old woman who presented with pressure symptoms, incidentally diagnosed with a vaginal mass and clinically suspected as vaginal myoma. This case was diagnosed based on histomorphology and immunohistochemistry and underwent surgical treatment. This tumor was a diagnostic dilemma for other benign and aggressive spindle cell tumors in the same area, such as endometrial stromal nodule, spindle cell lipoma, solitary fibrous tumors, and mixed tumors of the vagina.

Keywords: *Cellular angiofibroma, endometrial stromal nodule, mesenchymal tumor, solitary fibrous tumor*

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Introduction

Cellular angiofibroma (CA), a rare benign soft-tissue tumor that can affect both men and women, although it is most found in middle-aged women. It presents as a benign mass in the distal genital area, often without symptoms, typically occurring in the fourth through eighth decades of life, with a median age in the 40s.[1,2] Nucci *et al*. first described it in 1997.[2]

The pathogenesis of angiofibroma remains unknown, but several theories regarding its formation exist, with the angiogenic and histogenetic theories being the most widely accepted. A few studies have explored the role of sex hormones, genetics, and increased intracellular reactive oxygen species.[4]

Treatment for this tumor involves complete local excision, and recurrence has been described in only one case.[5] While it is not generally an aggressive tumor of mesenchymal origin, there have been reports of cases displaying aggressive behavior.[6-8]

Case Report

A 40-year-old female was incidentally diagnosed with a vaginal mass. She had mentioned a slight feeling of pressure in the vagina before discovering the mass but denied any pain or abdominal swelling. The patient had no remarkable medical or surgical history. She underwent a cervical cytology examination followed by complete resection of the mass, which was sent for histopathological examination.

Macroscopically, the mass was partly encapsulated and partly distorted, globular, with dimensions of 4 cm × 3 cm × 3.5 cm. The cut surface was homogeneously gray-white, without any evidence of necrosis, hemorrhage, or calcification [Figure 1].

On microscopy, tissue with stratified squamous lining along with an underlying well-circumscribed lesion composed of plump to oval to short spindled cells with variable cellularity and prominent vessels with hyalinization. The plump spindle cells were arranged in intersecting fascicles, sheets, storiform patterns, and in peritheliomatous arrangement, with areas of hyalinization. Individual cells had indistinct cell borders, moderate eosinophilic to clear cytoplasm,

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plump oval to mildly elongated nuclei with blunt ends, fine chromatin, and inconspicuous nucleoli. Areas of hyalinization were seen within the lesion. Many arterioles and medium vessels were interspersed within the lesion, with hyalinized vessels and focal whorling of the lesional cells around the vessels. There were no morphological epithelial islands/nests/increase in mitosis noted [Figure 2].

On immunohistochemistry, the lesional cells were positive for CD34, focally positive for smooth muscle actin (SMA), desmin, caldesmon, Pan CK, and CD10. Focally positive for estrogen receptor (ER) and progesterone receptor (PgR) was also seen [Figure 3]. The lesional cells were negative for EMA, cyclin D1, and CD117. IHC for STAT6 was not performed.

The diagnosis of CA was made based on morphological and immunohistochemical features. The postoperative period was uneventful.

Discussion

CA is a relatively recently described rare benign soft-tissue tumor. This tumor occurs predominantly in the vulvoperineal region. It affects a benign nodular mass of nearly 3 cm with or without symptoms in the fourth through eighth decades of life, with a median age in the 40s.[1,3]

In addition, CA cases were reported in extragenital regions such as the knee, anus, retroperitoneum, and oral mucosa.[9] Clinically and morphologically, it can resemble other benign tumors, including leiomyoma, solitary fibrous tumor, spindle cell lipoma, hemangiopericytoma, and perineurioma.[9]

Very few cases have been reported in the literature to date. The literature review is enlisted in Table 1.

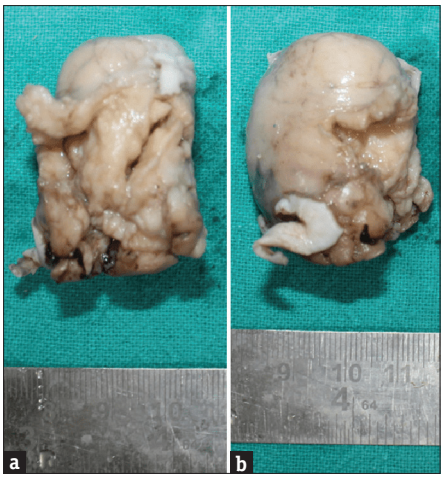

Figure 1: Gross morphology shows partly encapsulated and partly distorted globular mass lesion. (a: One surface, b: Other surface)

Diagnosing CA can cause challenges due to its morphological similarities with other soft-tissue tumors such as endometrial stroma nodules, solitary fibrous tumors, mixed tumors of the vagina, and hemangiopericytoma. Therefore, it was necessary to consider other differential diagnoses.

In this case, the morphology was of a benign spindle mesenchymal lesion, with differential diagnoses of solitary fibrous tumor and cellular leiomyoma. Immunohistochemical expression of markers such as CD34, focal expression of desmin, and SMA leads to a conclusion of CA. Significant differences exist in the reported immunoprofile for this tumor, and more research is needed to resolve some of these discrepancies. Vimentin has consistently shown strong diffuse reactivity, and CD34 expression has been reported in nearly half of the cases.[5,13,14] Actin and desmin expression are found in 20% and 10% of tumors, respectively. Estrogen and PgR proteins have been found in up to 50% of cases.[1]

Aggressive angiomyxoma is sparsely cellular with infiltrative margins. Its diffuse desmin positivity and negative CD34 expression aid in differentiation from other aggressive tumors like solitary fibrous tumors.[8] These rare tumors require evaluation beyond morphology to distinguish them from other tumors such as endometrial stromal nodules, mixed tumors of the vagina, and more locally aggressive tumors such as aggressive angiomyxoma and solitary fibrous tumors.[3,8] Furthermore, it was found that local recurrence is very infrequent.[15]

Hence, these rare tumors are to be further evaluated beyond histomorphology to distinguish them from

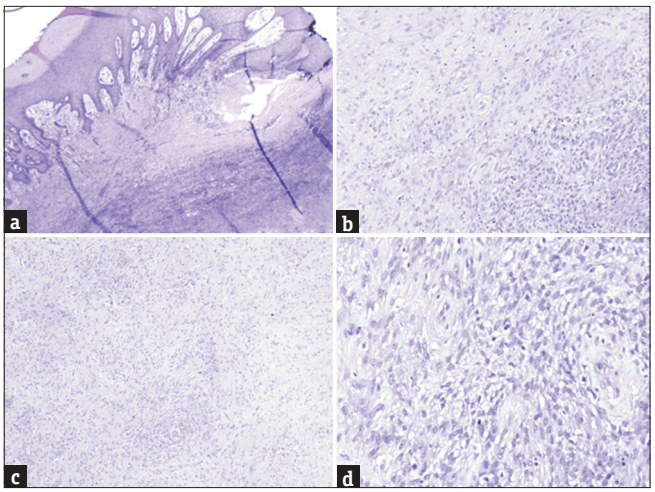

Figure 2: Micro pictograph shows circumscribed spindle cell lesion composed of plump to oval to short spindled cells with variable cellularity and prominent vessels with hyalinization. Areas with medium hyalinized vessels were interspersed within the lesion and focal whorling seen. (H and E, a: ×40, b: ×200, c: ×100 and d: ×400)

Table 1: Summary of literature review: Authors and key findings

Author and publishing year	Literature review
Nucci *et al*., 1997[3]	First described CA
Dargent *et al*., 2003[4]	Reported a case of CA
Iwasa and Fletcher, 2004[5]	Analysis of 51 cases of CA
Kairi-Vassilatou *et al*., 2011[6]	Reported a case of angiofibroblastoma of the vulva
Ahmadnia *et al*., 2014[7]	Reported a case of angiofibroma of the vulva
Magro *et al*., 2012[8]	Reported a case of vulvovaginal angiomyofibroblastomas
Magro *et al*., 2021[9]	Reported case series of aggressive angiofibroma
Bharti *et al*., 2014[2]	Reported a case of CA
Santiago *et al*., 2017[10]	A case report of CA of the vulva
Arega *et al*., 2019[11]	Reported a case of angiofibroma of the vagina
Cordaro *et al*., 2021[12]	Reported three cases of metachronous 13q14LOH mesenchymal tumors

CA: Cellular angiofibroma

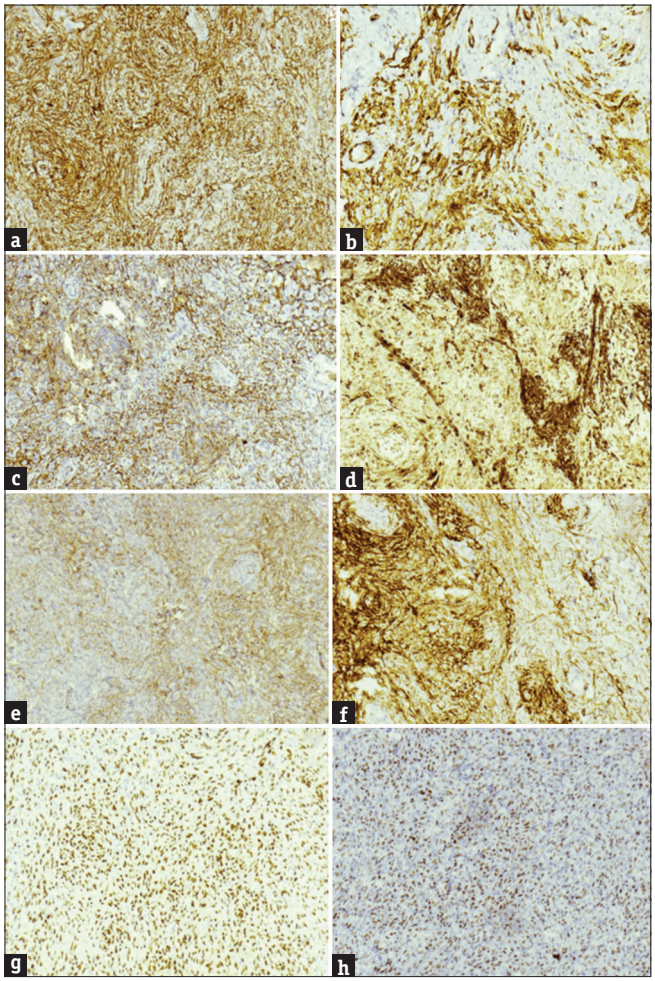

Figure 3: On immunohistochemistry, cells show positivity for CD34 (a), focally positive for SMA (b), panCK (c), Desmin (d), Caldesmon H (e), CD10 (f). Focally positive for estrogen receptor (g) and progesterone receptor (h)

more locally aggressive tumors such as aggressive angiomyxoma and various other mesenchymal lesions.[3,16] So far, cases with atypical features or sarcomatous transformation have not shown any signs of recurrence or metastasis.[15,17,18]

Conclusion

CA is a rare vulvar tumor that should be considered in assessing vulvar masses, especially in middle-aged and elderly females. It is crucial to distinguish these tumors from others in the same area. Pathologists should maintain a wide range of potential diagnoses in mind when dealing with vulvovaginal lesions, as numerous possibilities exist in this region. Surgical resection is the primary mode of treatment for this condition.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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