Case Report

Angiomyofibroblastoma of the Vulva: A Rare Case Report with Brief Review of Literature

Prita Pradhan, Nageswar Sahu, Kanakalata Dash, Pranati Misra, Urmila Senapati, Sudhanshu Kumar Rath[1]

Departments of Pathology and [1]Obstetrics and Gynaecology, Kalinga Institute of Medical Sciences, Bhubaneswar, Odisha, India

Abstract

Angiomyofibroblastoma is an unusual tumor of pelvic and vulval region. It is often diagnosed in the middle-aged women. Although benign owing to its location and morphology, it has a few benign and malignant mimics. Here, we present such a case where a 42-year-old female presented with a vulval mass.

Keywords: *Angiomyxoma, blood vessel, CD34, vimentin*

Introduction

Angiomyofibroblastoma is a rare tumor of vulval region. It is often diagnosed in the middle-aged women.[1] It is benign but owing to its location and morphology, it has a few benign and malignant mimics posing a diagnostic challenge.[2] Here, we present such a case where a 42-year-old female presented with a vulval mass.

Case Report

A 42-year-old female presented with swelling over right labia majora. There was no antecedent history of diabetes or hypertension. On examination, there is a well-defined, firm and nontender mass in the right labia measuring 4 cm × 3 cm × 3 cm. Based on the clinical findings, the differential diagnoses of lipoma and leiomyoma were considered. Incision and enucleation were performed.

Grossly, it was a well-circumscribed, globular gray-white, solid, firm mass measuring 4.8 cm × 3.3 cm × 2.5 cm [Figure 1a and b]. Sections show a well-encapsulated tumor containing alternating areas of hypercellular and hypocellularity [Figure 2a and b]. The hypocellular areas show stromal edema. There are abundant but irregularly distributed capillary sized blood vessels surrounded by aggregates of plump spindled stromal cells [Figure 2c]. Some stromal cells appear larger with mild nuclear atypia and mitosis <1/10 high power fields [Figure 2d]. Based on these features, the possibility of a benign myofibroblastic tumor was considered, suggestive of angiomyofibroblastoma. Immunohistochemistry was performed which showed positivity for CD34 in blood vessels and vimentin in the spindle cells [Figure 3], thereby confirming the diagnosis of angiomyofibroblastoma.

After the enucleation, the patient had an uneventful recovery and is disease free on follow-up for the past 1 year.

Discussion

Angiomyofibroblastoma an uncommon vulval tumor, most frequently encountered in the middle-aged women.[1] It is benign in nature but mimics a more aggressive tumor of the same location, aggressive angiomyxoma.[2] It is a tumor that is not very well characterized. Due to their slow growing and painless nature, Bartholin's cyst is the most frequent clinical impression though it can be confused with inguinal hernia or mesenchymal tumors, such as lipoma, leiomyoma, and liposarcomas.[1] Most of these tumors are grossly well circumscribed, homogeneous, tan to gray, tan mucofibroid to rubbery in consistency and range in size from 0.5 to 1 cm.[1-3] Rarely, they present as a pedunculated mass.[4] Radiologically, they appear as soft-tissue lesions with mixed echogenicity.[4] Microscopically, they classically show alternating hypocellular and hypercellular areas having entrapped scattered capillary sized blood vessels admixed with spindled stromal cells. The stromal cells have

Address for correspondence: Dr. Nageswar Sahu, Department of Pathology, Kalinga Institute of Medical Sciences, Bhubaneswar - 751 024, Odisha, India.
E-mail: nageswar.sahu@yahoo.in

How to cite this article: Pradhan P, Sahu N, Dash K, Misra P, Senapati U, Rath SK. Angiomyofibroblastoma of the vulva: A rare case report with brief review of literature. J Mid-life Health 2019;10:105-7.

Access this article online

Quick Response Code:

Website: www.jmidlifehealth.org

DOI: 10.4103/jmh.JMH_156_18

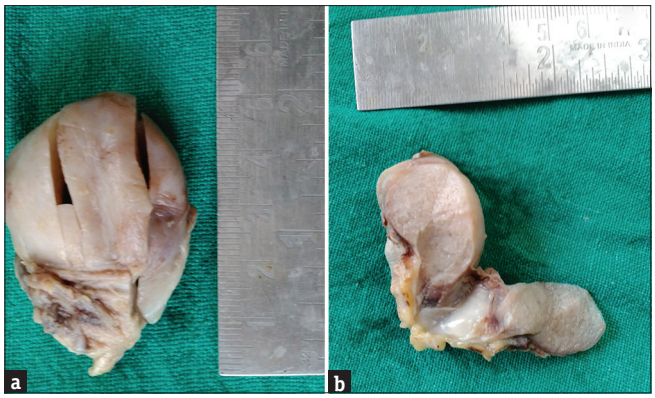

Figure 1: (a) Gross specimen of the enucleated vulval mass which was well-circumscribed, globular on the outer surface. (b) Cut surface of the mass is solid, gray-white firm measuring 4.8 cm × 3.3 cm × 2.5 cm

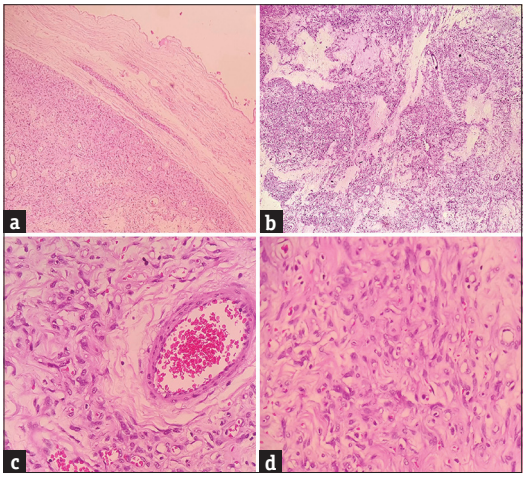

Figure 2: (a) Section from the tumor shows well-circumscribed mass having sharp circumscription (H and E, ×40). (b) Section from the tumor on scanner view showing alternate hyper- and hypo-cellular areas, the latter showing stromal edema. (c) Section from the hypercellular areas showing plump spindle cells showing a perivascular aggregates. (d) Section from the tumor showing stromal cells with mild nuclear enlargement, atypia and mitosis <1/10 high power fields

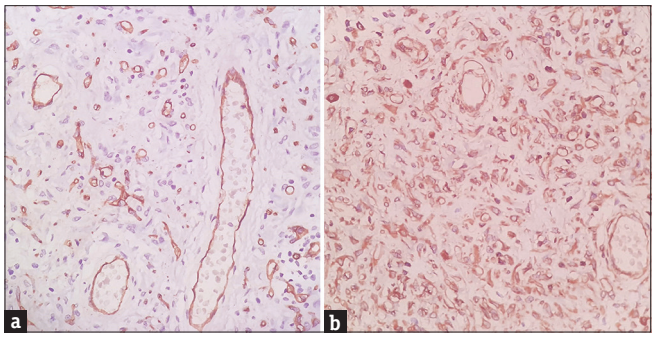

Figure 3: Immunohistochemistry showing CD34 positive in blood vessels (a) along with vimentin positivity in the intervening plump spindle cells (b)

a tendency for perivascular clustering.[1] Their nuclei showed minimal atypia with sparse to absent mitotic figures.[3] Multinucleated giant cells, plasmacytoid and epithelioid cells are also seen.[5]

On immunohistochemistry, the vessel shows positivity for the conventional markers for CD34. Stromal cells are vimentin and desmin positivity reactive for vimentin and desmin, variable positivity for smooth muscle actin, estrogen and/or progesterone receptors and negative S100 and cytokeratin.[1,3] This indicates myofibroblastic origin but owing to hormone receptor positivity; it likely arises from hormone responsive mesenchymal cells.[1,3]

On morphology, it can be mistaken for aggressive angiomyxoma based on its characteristic location, vascularity, and stromal cells. Angiomyofibroblastoma needs to be differentiated from its aggressive angiomyxoma as the latter is nonmetastasizing locally invasive recurring mesenchymal tumor.[2] The key features are sharp circumscription, cellular nature, and prominent capillary sized blood vessels with or without perivascular sclerosis admixed with stromal cells and absence of red cell extravasation.[3] Unlike angiomyxoma, there is no pervascular hyalinization or stromal mucin. Aggressive angiomyxoma positive for vimentin and negative for desmin.[6] Cellular angiofibromas show more thick-walled blood vessel and are negative for hormone receptors.[7] Other lesions of myofibroblastic differentiation like inflammatory myofibroblastic tumor show a distinct component of inflammatory cells.[8]

They are cured by simple excision and rarely recur.[2] Sarcomatous transformation rarely occurs.[9]

Conclusion

Angiomyofibroblastoma is an uncommon, benign tumor which occurs in the vulvovaginal region with benign and malignant mimics. Correlating the radiological data with characteristic histomorphologic and immunohistochemical findings help in making the diagnosis.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Seo JW, Lee KA, Yoon NR, Lee JW, Kim BG, Bae DS, *et al.*

Angiomyofibroblastoma of the vulva. Obstet Gynecol Sci 2013;56:349-51.
2. Nasu K, Fujisawa K, Takai N, Miyakawa I. Angiomyofibroblastoma of the vulva. Int J Gynecol Cancer 2002;12:228-31.
3. Fletcher CD, Tsang WY, Fisher C, Lee KC, Chan JK. Angiomyofibroblastoma of the vulva. A benign neoplasm distinct from aggressive angiomyxoma. Am J Surg Pathol 1992;16:373-82.
4. Giannella L, Costantini M, Mfuta K, Cavazza A, Cerami LB, Gardini G, *et al*. Pedunculated angiomyofibroblastoma of the vulva: Case report and review of the literature. Case Rep Med 2011;2011:893261.
5. McCluggage WG, White RG. Angiomyofibroblastoma of the vagina. J Clin Pathol 2000;53:803.
6. Das BP, Baruah D, Medhi KB, Talukder B. An aggressive angiomyxoma of vulva – A rare entity – A case report. J Midlife Health 2016;7:140-3.
7. Bharti JN, Dey B, Raj VG, Arora P. Cellular angiofibroma of vagina presenting with secondary infertility. J Reprod Infertil 2014;15:165-7.
8. Qiu P, Wang Z, Li Y, Cui G. Giant pelvic angiomyofibroblastoma: Case report and literature review. Diagn Pathol 2014;9:106.
9. Nielsen GP, Young RH, Dickersin GR, Rosenberg AE. Angiomyofibroblastoma of the vulva with sarcomatous transformation ("angiomyofibrosarcoma"). Am J Surg Pathol 1997;21:1104-8.