CASE REPORT

Angiomyofibroblastoma Presenting as Labia Minora Cyst

Kafil Akhtar[1], Binjul Juneja[1], Mohd Talha[1], Zehra Mohsin[2]

[1]Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, Uttar Pradesh, India, [2]Department of Obstetrics and Gynaecology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, Uttar Pradesh, India

ABSTRACT

Angiomyofibroblastoma (AMF) is an uncommon soft tissue tumor which occurs principally but not exclusively in the vulvovaginal region of women in their reproductive years. Rarely, in males, the tumor may occur in the scrotum or par testicular tissues with the age ranging from 40 to 80 years. AMF has a diverse histologic and immune histochemical profile. It is a circumscribed edematous slow-growing mesenchymal tumor. Our case report presents a 25-year-old female with a cystic lesion over the labia minor, which was clinically mistaken to be a Bartholin's cyst. In general, AMF is benign, with no local recurrence or metastasis. Hence, wide surgical excision is the sufficient treatment modality. We have also discussed the possible differential diagnosis and their differentiating features on the basis of histology and immune histochemistry.

Key words: Angiomyofibroblastoma, immunohistochemistry, labia minora

INTRODUCTION

Angiomyofibroblastoma (AMF) is a benign mesenchymal tumor, with two major components, prominent blood vessels and stromal cells. Frequent plump stromal cells of epithelioid and spindle cell types are arranged around numerous blood vessels.[1] Typically, the tumor presents as a small slowly enlarging and painless mass. These lesions can be large, painful, and/or pedunculated.[2,3] AMF is usually well-circumscribed unencapsulated mass, of size <5 cm involving the external genitalia and perineal soft tissue. Frequently, fat in the lesion may occasionally be very prominent, thence known as "lipomatous variant." The differentials of AMF are fibroepithelial stromal polyps, cellular angiofibroma, aggressive angiomyxoma, myofibroblastoma, leiomyomatosis, and Bartholin's gland cyst.[4]

CASE REPORT

A 25-year-old married female presented in the obstetrics and gynecology clinics with a cystic swelling around the labia minora, which had gradually increased in size during the past 2 months. On examination, the mass was cystic, 2 cm × 1.5 cm in size, painless with regular margins. Medical history was not significant. Internal genital organ examination and menstrual history were normal. She denied any history of sexually transmitted disease or gynecology-related surgery. Complete removal of the cystic swelling was performed with clear cut margins. Histopathological examination showed alternate areas of hypercellular and hypocellular zones with hyperkeratotic stratified squamous epithelial covering with focal acanthosis, loose stroma composed of plump spindle cells, and occasional bizarre cells along with irregularly distributed dilated non-hyalinized thin-walled blood vessels [Figures 1 and 2]. Immunohistochemically, the stromal cells were reactive for vimentin and desmin but negative for cytokeratin, smooth muscle actin, and S-100 protein. Based on these features, the patient was diagnosed as AMF. On follow-up after 6 months, our patient is well with no sign of metastasis or recurrence.

Address for correspondence:
Dr. Kafil Akhtar, Department of Pathology, Jawaharlal Nehru Medical College, Aligarh Muslim University, Aligarh, Uttar Pradesh, India. E-mail: drkafilakhtar@gmail.com

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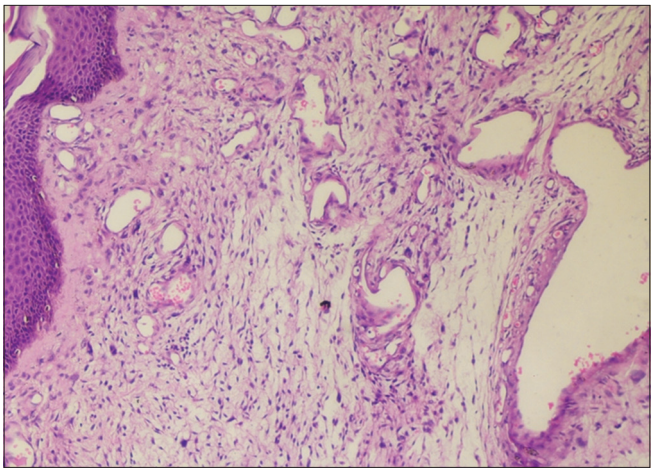

Figure 1: Histopathological examination shows alternate areas of hypercellular and hypocellular zones with hyperkeratotic stratified squamous epithelial covering with focal acanthosis, loose stroma composed of plump spindle cells, and occasional bizarre cells along with irregularly distributed dilated non-hyalinized thin-walled blood vessels. Hematoxylin and eosin ×10

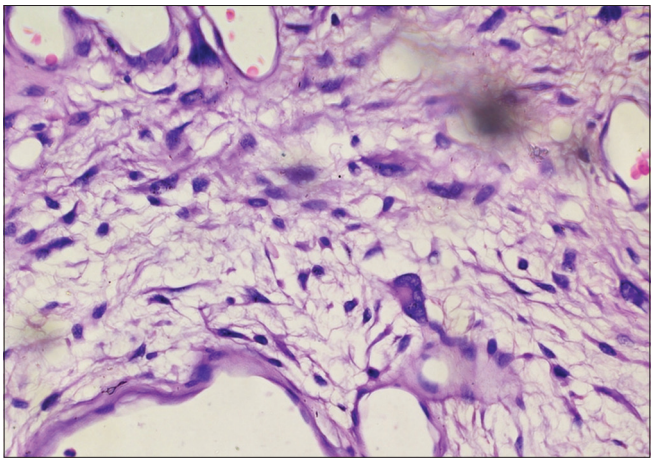

Figure 2: Histopathological examination shows stratified squamous epithelial covering with loose stroma composed of plump spindle cells, occasional bizarre cells along with irregularly distributed dilated non-hyalinized thin0walled blood vessels. Hematoxylin and eosin ×40

DISCUSSION

The first case of AMF was reported by Fletcher *et al*., in 1992.[5] It is a benign myofibroblastic tumor common in females of reproductive age. Around 10.0% of patients are postmenopausal.[1] It is rare in males and presents as scrotal or par testicular mass. AMF is slow-growing, painless, well-circumscribed pseudoencapsulated tumor, usually of size <5 cm. Histological characterized by alternating hypercellular and hypocellular areas admixed with small blood vessels. Spindle and plump stromal cells aggregate around the vessels. There may be a component of mature adipose tissue, which when abundant, the tumor is referred to as the lipomatous variant.[6] If the vessels are not prominent; the term myofibroblastoma (without the prefix "angio") is used. The tumor cells are immune reactive for vimentin, desmin, and CD34, but negative for actin and keratin.[7,8] It is hard to escape from the suspicion that this tumor is histogenetically related to the other tumors. Cellular angiofibroma is more than 5 cm in size which is a distinguishing feature. It is not circumscribed and shows moderate cellularity with large thick hyalinized walled vessels. The spindle cells may stain for actin and desmin but are negative for S-100 protein, factor VIII-related antigen, carcinoembryonic antigen, and keratin. Removal of these neoplasms is sometimes difficult due to their infiltrative nature. They tend to recur, sometimes repeatedly. Fibroepithelial stromal polyp is a poorly circumscribed exophytic, submucosal growth with frequent bizarre stromal cells. These lesions typically do not grow larger than 5 cm in diameter and are found incidentally during routine gynecologic examinations.[9] The most characteristic feature is the presence of stellate and multinucleate stromal cells which are usually identified near the epithelial-stromal interface.[9] AMF has a higher cellularity and more numerous blood vessels than aggressive angiomyxoma, its major neoplastic contender which exhibits aggressive behavior and is treated differently from benign tumors.[2,4,6] Per vascular clustering of cells in angiomyofibroblastoma may suggest glomus tumor, but the later has uniform cellularity with very regular round cells. Clusters of epithelioid cells in AMF may suggest epithelioid leiomyoma. The variable immune staining patterns characterize and differentiate AMF from other mimickers. Although immune histochemical staining is indispensable in the work-up and diagnosis of AMF and other vulvovaginal tumors; few important diagnostic features of the lesion are size under 5 cm, restricted to external genitalia and perineal soft tissue, circumscribed with thin pseudocapsule, edematous background with prominent vascularity, and alternating hypercellular and hypocellular areas. Grading and staging are not applicable due to its low recurrence or metastasis. Therefore, simple total surgical excision of the lesion is the treatment of choice.

CONCLUSIONS

AMF of labia minora is a rare benign tumor. Due to its diverse histologic and immune histochemical profile, differentiation is indispensable from some aggressive tumors.

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