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Vulval Angiomyofibroblastoma - A Rare Case Report

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Abstract

Angiomyofibroblastoma is a benign soft tissue tumour arising from the mesenchymal cells. It is seen in young females most commonly arising in the vulval region. This tumour mimics other benign tumours of the vulva and is pathologically differentiated from Angiofibroma and AMM (Angiomyomyxoma) which follow a different treatment protocol. This tumour is rare in occurrence and is unusual in older females. Here we report a rare case of AFMB in an older woman who presented with a very slow-growing asymptomatic vulval mass involving the left labia majora.

Keywords: *Angiomyofibroblastoma, Benign, Soft Tissue Tumour, Vulva.*

Introduction

An uncommon benign mesenchymal tumour is called angiomyofibroblastoma (AMFB). Middle-aged women's vulvovaginal regions are the main site of AMFB [1-3]. It commonly arises in the vulva but can also occur in the perineum, vagina, retroperitoneum and inguinoscrotal region, scrotum, and occasionally spermatic cord in men [13]. Diagnosis is made based on pathological findings and immunohistochemistry. Angiofibroma and Angiomyxoma are differential diagnoses [19]. Sometimes, AMFB is misdiagnosed as a Bartholin gland cyst due to its slow-growing nature and possible formation of a pedunculated mass [7]. The importance of exact diagnosis is emphasized in this article as tumours like AMM require more aggressive resection with wide margins. This is a case report of a 75-year-old woman who has AMFB of the vulva, on her left labia majora.

Methodology

A 75-year-old female presented to the outpatient department with a slow-growing, painless vulval mass, on the left labia majora that had begun a few months prior. She initially disregarded the lump despite its increase in size as she had no symptoms. Gradually as the lump increased in size she had a dragging sensation and feeling of discomfort at a later stage. Therefore she sought medical help for the same. Her general examination was unremarkable. Upon physical examination, there was a swelling of 2x2 cm on the left labia majora (figure 1). It was a well-defined hard mass with tense and stretched-out skin. There was no tenderness and no induration of the surrounding skin. There were no other swellings in the perineum. The vaginal examination was normal. Inguinal lymph nodes were not palpable. However ultrasound pelvis and bilateral inguinal region was done and pelvic, inguinal lymphadenopathy and pelvic mass were ruled out. A clinical diagnosis of Bartholin's cyst was made. The decision was made for simple excision of the tumour. Routine investigations for surgery were done which were found to be normal. Excision of the lesion was carried out under local anaesthesia (figure 1) and the specimen was sent for the

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Frozen section. The initial report came to be a benign lesion with all margins and depth free of tumour cells. Hence closure was done and the patient was managed as a daycare. She was followed up regularly for post-operative care, suture removal and to check for any recurrences. The pathology report showed a polypoidal mass lined superficially by squamous epithelium. The underlying dermis showed well-circumscribed unencapsulated neoplasm composed of ectatic blood vessels of varying sizes surrounded by hyalinized fibrous tissue with occasional areas showing plump spindle cells (Figure 2&3). These features were suggestive of benign lesions with a differential diagnosis of cellular angiofibroma and AFMB. To confirm the diagnosis Special staining of Desmin and Vimentin was done. In our case, the tumour stained positive for Vimentin (figure 4) and negative for Desmin (figure 5). The diagnosis of AFMB was confirmed.

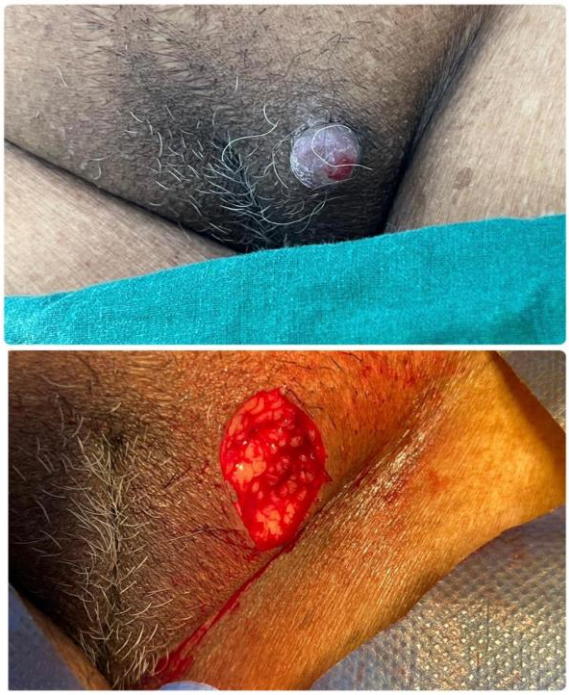

Figure 1. Showing Excision and Biopsy of Lesion in Left Labia Majora

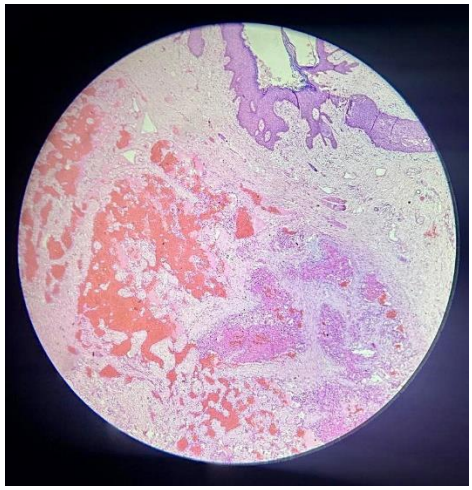

Figure 2

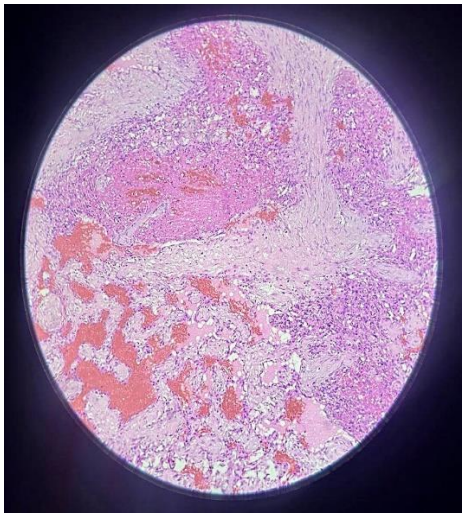

Figure 3

Figures 2 and 3 Showing Histopathological Images of "Skin with an underlying unencapsulated tumour composed of abundant irregularly distributed blood vessels surrounded by myxoid and edematous stroma. The individual tumour cells are bland spindle-shaped to epitheloid, arranged in nests and short intersecting fascicles"

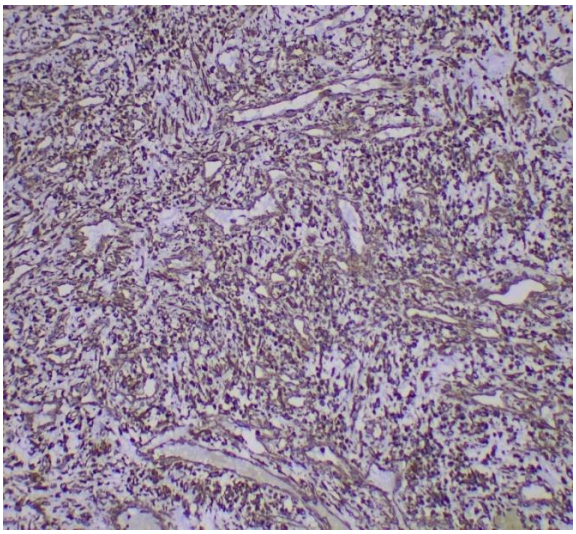

Figure 4. Showing Positivity for VIMENTIN

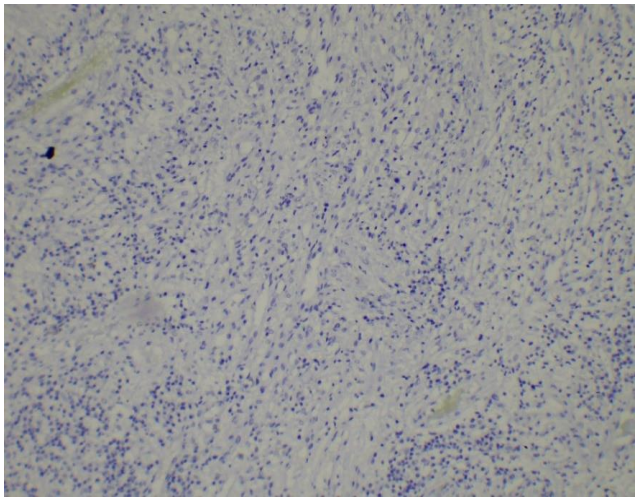

Figure 5. Showing negative for DESMIN

Results

The patient recovered completely with healing of the surgical site. She was followed up for 2 years. There was no recurrence of the tumour at the operated site or in the surrounding vulval region. The patient is advised to follow up regularly.

Discussion

The most commonly encountered tumours in the vulva are hemangioma, lipoma, fibroma, and leiomyoma [14]. Angiomyofibroblastoma is a rare benign tumour arising from the mesenchymal cells [20], Fletcher et al [8] 1992 described 10 such cases in the vulval region and made AMFB widely known. There were just 71 AMFB cases reported in English literature between 1999 and 2013 [13]. In 2015, Wolf et al. described 125 cases of female AMF in their review of literature, and most of the cases were either vulvar or vaginal in origin [15]. These tumours are very slow-growing tumours with patients seeking medical help at a later stage as in our case. This tumour mostly arises in young and middle-aged females [4] in their reproductive age group, unlike in our case, who is in her late seventies which is not so usual. Some studies have described females typically aged between 40 and 70 years old presented with slow-growing painless tumours with a duration of from a few weeks to 13 years old [5]. These tumours are most commonly found in the vulva vaginal region and perineum. The scrotum and inguinal region in men is a rare occurrence [8,9].

All published findings indicate that patients with AMFB have a benign clinical result when it comes to biologic behavior Sarcomatous change and tumour recurrence is a rare possibility. There has only been one documented case that experienced a sarcomatous change [5] and no subsequent recurrence. Clinically, these tumours are mistaken for other mesenchymal tumours like lipoma, angiomyxoma, leiomyoma, or Bartholin gland cysts [11]. Though the initial differential diagnosis of angiofibroma was made based on the pathological report, angiofibromas are a rare occurrence in the vulval region.

AFMBs are usually well-defined, firm, and have a rubbery texture. Their sizes normally vary from 5mm to 12 cm. The majority of tumours have a pink hue [5]. On microscopic examination, alternating hyper-cellular & hypo-cellular [17] edematous areas [4], myxoid stroma with many blood vessels and plump stromal cells are visible [16]. Tumor cells typically gather near blood vessels and exhibit nuclear atypicality or mitotic features infrequently [5]. These tumours are categorized as the “lipomatous” variety of AMFB when there are occasionally adipocytes dispersed throughout the mass. The well-defined boundaries are a feature of AMFB [7] while aggressive angiomyxomas are invasive. The stromal cells in AMFB are plump and condense the perivascular region [12] as in our case. The majority of the time, vimentin and desmin are stained by these tumours. In addition, the tumour cells express muscle actin inconsistently and have positive progesterone and/or estrogen receptors [5]. In our case, the tumour stained positive for vimentin which is not a feature of Angiofibroma or Angiomyxoma [6]. Therefore the diagnosis of AFMB was made.

Treatment requires simple excision with clear margins as in our case. As this is a benign lesion, the prognosis is very good and there is almost no chance of recurrence any occasional recurrence is due to incomplete surgical excision which can simply be addressed by re-excision [13,18]. In our case, we followed up with the patient for about two years. Our patient didn’t have any local recurrences as expected. We are regularly following up with the patient on the phone as she could not visit the hospital and she claims that there was no recurrence and forgot about the lesion and the surgery.

Conclusion

The vulvar angiomyofibroblastoma(AMFB) is an uncommon rare progressive growing benign tumour with delayed presentation. They mimic other benign tumours like Bartholins cyst, lipoma and AAM. Surgical excision and pathological examination is the only way to confirm the diagnosis. Pathologically this tumour should be differentiated from AAM which is relatively aggressive compared to AFMB and requires wide local excision. Though recurrence and malignant transformation in AFMB is rare, it cannot be completely ruled out and patients followed up for the same. More research and awareness among medical personnel would fill the gaps in understanding this rare entity.

Acknowledgement

Without the involvement and support of several people, many of whose names may not be listed, this project would not have been able to be completed. We sincerely thank them for their contributions and acknowledge them.

Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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