ORIGINAL RESEARCH PAPER

Gynaecology

ANGIOFIBROBLASTOMA OF THE VULVA: A RARE PELVIC PATHOLOGY

KEY WORDS: angiomyofibroblastoma, vulva, postmenopausal

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ABSTRACT

Angiomyofibroblastoma is a rare mesenchymal tumour mostly occurring in the lower genital region of premenopausal women. They are usually benign in nature and composed of two components namely blood vessels and stromal cells. This is a rare case report of an angiomyofibroblastoma occurring in a 74 year old postmenopausal woman. Her only presenting symptom was a painless slow growing lump in the vulval region. This patient underwent a wide local excision with vulvar reconstruction.

INTRODUCTION

Angiomyofibroblastoma (AMF) is a rare, benign, genital tract typically affecting women in their reproductive age group. It belongs to a diverse group of genital mesenchymal tumors1. The vulva, perineum, vagina and the uterine cervixare common sites of predilection of this lesion2. Though uncommon, AMF may also involve the lower genital tract and the inguinoscrotal regions in male. Its occurrence in the pelvis and the retroperitoneum has also been reported 3.

It is clinically often confused with a Bartholin's cyst, fibromyxoid sarcoma, hydrocele of the Canal of Nuck, and most importantly aggressive angiomyxoma2.

CASE REPORT

A 74 year old multiparous, postmenopausal woman presented with a slow growing palpable lump in her external genitalia. She first noticed the mass 6 years prior to the outpatient visit. Initially the mass was small in size and had gradually progressed over time. The mass had remained painless through its course of progression. She denied a history of discharge per vaginum, weight loss or fever. She had normal bowel and bladder habits.

In the past, the patient had undergone 5 vaginal deliveries and an abdominal hysterectomy due to presence of multiple fibroids. She also suffered from hypertension land diabetes since 30 years and was on regular medication. No other medical or surgical history was elicited.

An examination following informed consent revealed a large swelling arising from the right labia minora. The approximate dimensions of the lesions were 8 x 7 x 6 cm (AP x TR x CC) as shown in fig 1. The swelling was firm, fixed and not tender on palpation. Its consistency was intermediate to solid and cystic with no appreciable evidence of surface ulceration or slough. There was no palpable pelvic lymphadenopathy. Per speculum examination revealed an atrophic vaginal vault. A clinical diagnosis of a Bartholin's tumour was made in the background of clinical suspicion of possible carcinoma of the vulva.

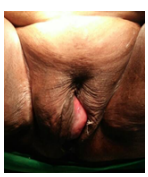

fig 1

The patient was admitted for further investigations and management. Besides laboratory workups, an ultrasound of the pelvis was prescribed in order to evaluate lesion. The ultrasonogram of the labia revealed the presence of a lesion of heterogeneous echotexture with significant evidence of vascularity on Doppler interrogation. No evidence to suggest calcification or tissue breakdown was detected on the ultrasound. Further, a contrast enhanced MRI was requested for preoperative planning and staging of the disease. It revealed a 6.4 x 4 x 6.5 cm sized (AP x ML x CC) lesion (fig 2a) with signal and enhancement heterogeneity. Along its anteromedial aspect , the lesion was seen exerting mass effect in the form of displacement of the urethra with maintained intervening fat planes as shown in fig 2b. No pelvic and inguinal lymphadenopathy was detected.

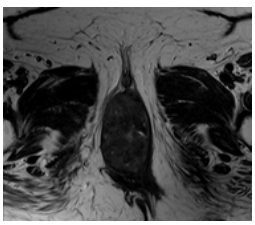

fig 2a.

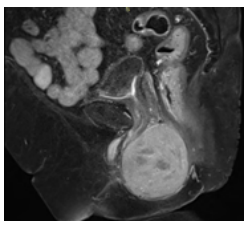

fig 2b.

Patient was posted for a wide local excision with vulval reconstruction following anaesthetic fitness clearance.

Intraoperatively, a frozen section of the specimen was obtained. No frank evidence of malignancy was detected. The tumor was excised en block and sent for further pathological evaluation. Surgical specimen was 8 x 7 x 6 cm as shown in fig 3a and 3b.

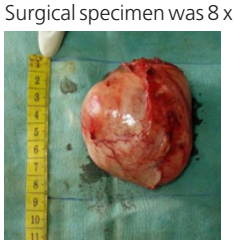

fig 3a.

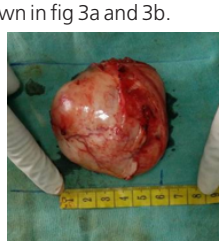

fig 3b.

Post-operative monitoring and wound care ensured an uneventful hospital stay and rapid recovery. Microscopic histopathological analysis of the specimen revealed a lesion composed of alternate areas of hypo and hypercellularity. Areas of stromal edema were also observed. The tumour cells were spindle shaped and arranged in fascicles with irregularly distributed thin walled vessels. There was no evidence of atypical, necrosis or malignancy consistent. (As shown in fig 4 and fig 5)

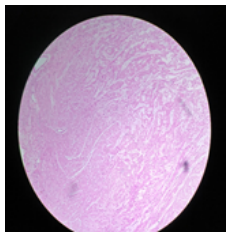

fig 4a

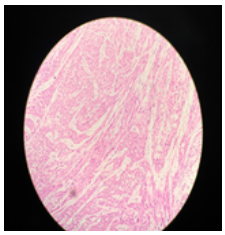

fig 4b

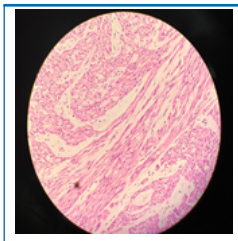

fig 4c

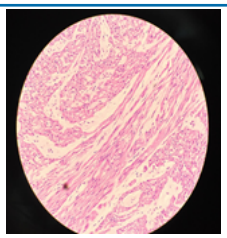

fig 4d.

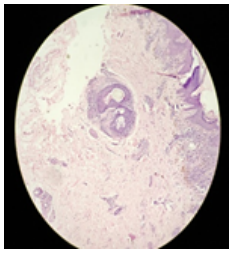

Fig 5a

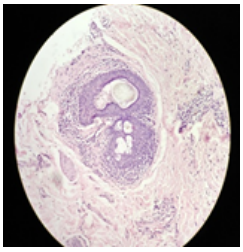

Fig 5b.

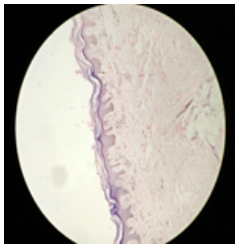

fig 5c

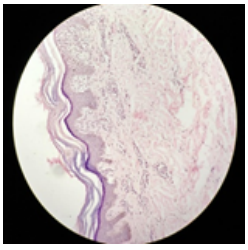

fig 5d.

These histopathological findings were consistent with a diagnosis of angiomyofibroblastoma of vulva.

DISCUSSION

Angiomyofibroblastoma is an uncommon benign tumor of mesenchymal origin. AMF essentially affects premenopausal women with a predilection to occur at the vulvovaginal region. Other rare sites of origin of this tumor have also been reported in the upper genital tract (uterus and fallopian tubes[4]). The tumour presents as a slow growing painless lump with an indolent course. Occasionally urinary complaints secondary to mass effect over the urinary tract may be amongst the other presenting symptoms. Albeit its benign nature, one case of malignant transformation has been reported by Nielsen et al [5].

Clinically, the tumour may be confused with other benign and malignant neoplasia of the vulval conditions. Clinical differentials include Bartholin's cyst, benign lipomas and carcinoma of the vulva. Though fibroepithelial stromal polyps and cellular angiofibromas may be encountered, these are clinically indistinguishable [6].

Pathology plays a crucial role in the diagnosis of this lesion. Microscopically, AMFs are composed of stellate or spindle shaped cells with bland nuclei and eosinophilic cytoplasm. The cells are arranged around capillary type blood vessels admixed with mature adipose tissue seen in alternating layers of hypercellular stroma and hypocellular edematous area. The tumor has high cellularity with well-formed blood vessels Among the various clinicopath ological differentials, Aggressive angiomyxoma (AAM) is of prime consideration. AAM is a malignant and locally invasive lesion with a tendency to recur following excision. Aggressive angiomyxoma and angiomyofibroblastoma mimic each other clinically with respect to age at presentation, location and symptomatology. At pathology, AAMs are characterised by abundant myxoid matrix, infiltrating surrounding tissue, irregular margins, low cellularity, thickened medium to large sized vessels and extravasation of red blood cells [7,8,9,10]. AAMs show a more distinctive area of myxoid degeneration than than AMFs[1,11]. Following surgical treatment, 30% of AAMs have been found to relapse within 2 years [1]. Immunohistochemistry shows that the tumors are positive for vimentin and desmin and variable expression for muscle actin and are positive for estrogen and/or progesterone receptors as well [12].

The important differential is a Cellular Angiofibroma (CA). CAs are tumors which also belong to the spectrum of genital mesenchymal tumours. Angiomyofibroblastoma, however, is distinguished from CA by the presence of round tumor cells grouped around vessels, and a lack of wispy collagen bundles and numerous thick-walled, hyalinized vessels.

The treatment of choice for AMF is simple total excision. The procedure is usually curative with almost no postsurgical incidence of recurrences or metastasis, thus confirming its benign nature[13].

CONCLUSION

The case presented in the manuscript describes an unusual tumor occurring in an uncommon age group. Pelvic AMF is an extremely rare but benign tumor of the genital tract. Its preoperative diagnosis and differentiation from other soft-tissue tumors are challenging owing to its low incidence and propensity to clinically mimic other conditions. The combination of imaging along with histological and immunohistochemistry confirms the diagnosis. The knowledge of the condition and simplicity of treatment is valuable in order to avoid unnecessary investigations thus alleviating possible anxiety and cosmetic deformities in patients especially those in the reproductive age group.

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