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Case Report

AGGRESSIVE ANGIOMYXOMA OF VULVA- A RARE ENTITY

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ABSTRACT

Aggressive angiomyxoma (AA) is a slow-growing vulvovaginal mesenchymal neoplasm affecting women of reproductive age and is associated with a high risk of local recurrence. Usually, it presents as a vulval polyp clinically and needs to be distinguished from other benign myxoid tumors with a low risk of local recurrence. It is diagnosed only on histopathology. The treatment of choice usually being wide local excision with tumour-free margins and occasionally hormonal manipulation. We present here a case of a 40-year-old female presenting with a large, fleshy, polypoidal mass on the right labia majora.

INTRODUCTION

AA was described by Steeper and Rosai in 1983. It is a rare, slowly growing, and benign tumour of mesenchymal origin with a predilection for the perineum , pelvis, vulva, vagina and urinary bladder in adult women of reproductive-age women [1]. Estrogen and progesterone receptors are commonly found in AA.[2] Thus, it is likely to grow during pregnancy and respond to hormonal manipulation. The term "aggressive" denotes its propensity for local aggression and recurrences after excision [3] so, appropriate management and long-term follow-up are necessary for proper diagnosis. Very few cases have also been described in men, usually involving the scrotum. Female to male ratio is 6:1 [2]. A 40-year-old female with a polypoid vulva lesion which turned out to be AA, and its clinicopathological characteristics of this rare entity are discussed in this case report.

CASE REPORT

A 40-year-old female was admitted to gynaecology ward with an polypoid mass on her right labia majora since two years. The mass was gradually increasing in size. On clinical examination a well-circumscribed polypoidal mass measuring approximately 15 × 10 × 6 cm was revealed. The lesion was non tender, soft and gelatinous in consistency, covered by normal appearing skin and no enlargement of inguinal lymphnodes was seen. There was no history of any vulval discharge or bleeding. Menstrual cycles were regular with a normal flow.

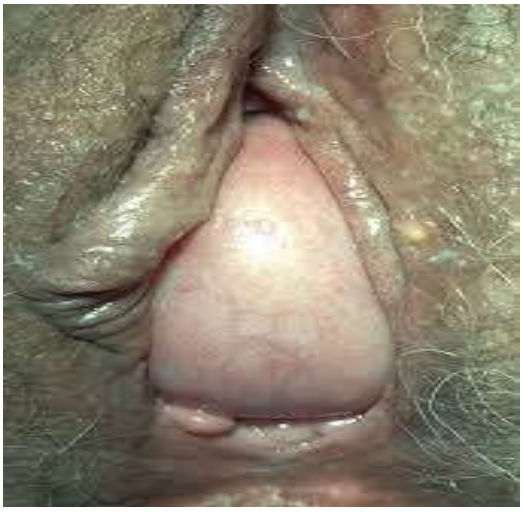

Fig 1 Clinical Photograph-Showing an Polypoidal mass near labia majora.

On gynaecological examination the cervix and vagina were unremarkable. Her laboratory investigations were within normal limits and ultrasonography of the abdomen was unremarkable. Ultrasonography of the perineum showed a large mass with peripheral vascularity, heterogenous hyperechoic areas, measuring approximately 16 × 10 × 6 cm, and magnetic

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resonance (MRI) of the pelvis confirmed these features. Fine needle aspiration cytology was inconclusive revealed only blood without cellular elements. Excisional biopsy of the lesion was planned considering a differential diagnosis of fibroepithelial polyp or a vulval fibroma. Specimen was sent to our department for histopathological study. Grossly the size of tumour was 15x10x5 cm.

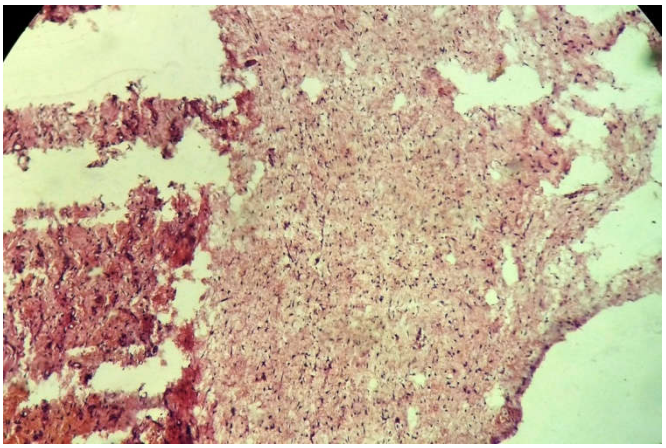

Fig 2 Photomicrograph scanner view (40x)-Hypocellular spindle cells in myxoid background.

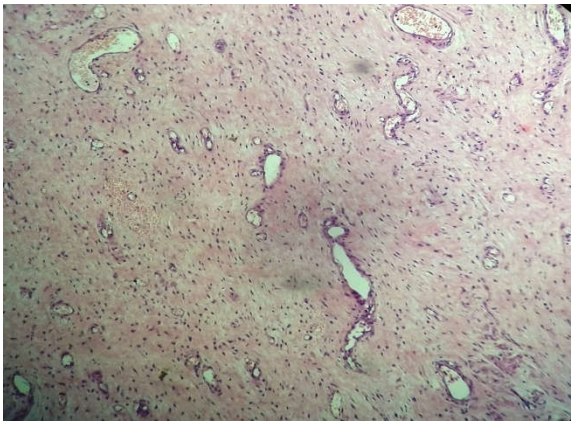

Fig 2 (b) Photomicrograph LP100X-showing spindle cells along with thin and thick walled blood vessels in myxoid background.

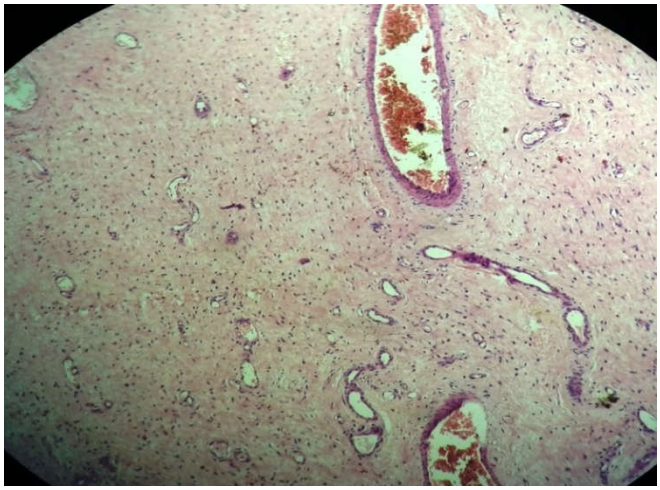

Fig 2 © Photomicrograph LP 400x- showing hypocellular spindle cells and thick walled blood vessels.

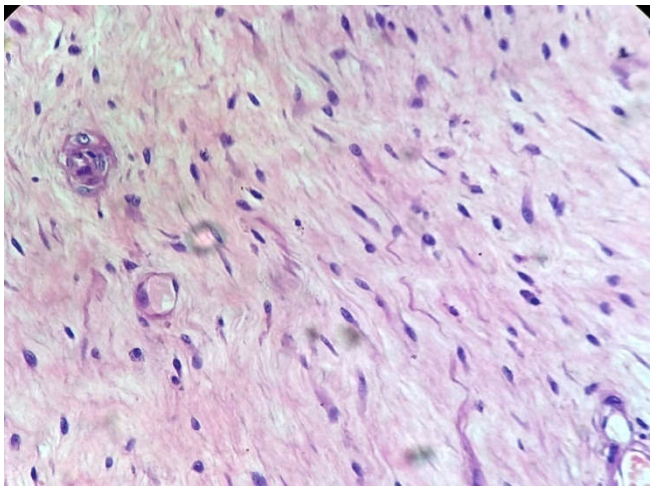

Fig 2(d) Photomicrograph-HP400x- scattered spindled to stellate-shaped cells with ill-define cytoplasm and small round to oval hyperchromatic nuclei with small centrally located nucleoli, embedded in a myxoid stroma.

The cut surface revealed a glistening, gelatinous, and soft homogeneous appearance. On microscopic examination the tumor was composed of spindle and stellate‑shaped cells embedded in a loose matrix with wavy collagen. The lesion contained thin-walled capillaries and thick-walled vascular channels. Neoplastic cells had relatively scant eosinophilic cytoplasm and their nuclei were ovoid, containing finely dispersed chromatin with one or two small eosinophilic nucleoli. There was no significant nuclear pleomorphism nor mitoses was observed. IHC was done which was positive for vimentin and CD34.Thus a diagnosis of AA was rendered.

DISCUSSION

Angiomyxomas are classified either as superficial (also called as cutaneous myxoma) or AA. Superficial angiomyxomas usually present in middle-aged adults as a single nodule or a polypoidal lesion in the head and neck region .They may occur in the setting of Carney complex [4] and may be clinically confused with skin tag or neurofibroma. On the other hand, Agressive angiomyxoma (AA) occurs almost exclusively in the pelvic and perineal regions of women of reproductive age. Usually, this tumor does not metastasize, but there are reports of multiple metastases in women treated initially by excision and ultimately succumbing to it.[5,6].

The main theory for AA pathogenesis is unclear. However this hormonally responsive tumor is believed to arise from specialized mesenchymal cells of the pelvic-perineal region or from the multipotent perivascular progenitor cells, which often display variable myofibroblastic and fibroblastic features.[7]. The latter is mainly supported by immunohistochemical expression of desmin and, in some cases, a smooth muscle actin along with desmin by tumour cells [8].

AA is molecularly part of the benign group of mesenchymal tumours showing multiple aberration region involvement [9, 10]. HMGIC expression in AA is of value in the histological differential diagnosis. Molecular studies have linked a consistent clonal aberration of the chromosome 12, in the region 12q13-15, associated with rearrangement of the HMGIC gene (high-mobility group protein isoform I-C) with AA.

On computed tomography (CT) scan, these tumors have a well-defined margin with attenuation less than that of the muscle. On MRI, these tumors show high signal intensity on T2-weighted images. The attenuation on CT and high signal intensity on MRI are likely to be related to the loose myxoid matrix and high water content of angiomyxoma.[11].

Clinically, AA may be misdiagnosed as Bartholin cyst, lipoma, labial cyst, Gartner duct cyst, or sarcoma. Fibro-epithelial stromal polyp, superficial angiomyxoma, angiomyofibroblastoma, cellular angiofibroma and smooth muscle tumors also need to be considered in the differential diagnoses of a polypoidal mass in the perineum. AA is an infiltrative tumor, whereas angiomyofibroblastoma is well circumscribed (this characteristic feature can also be identified on magnetic resonance imaging [MRI]). Also, AA has thick-walled vessels, which are less numerous than the thin-walled vessels in angiomyofibroblastoma.

The diagnosis of AA is opined in this case based on the distinct histopathological findings. Tumour is composed of widely scattered spindled to stellate-shaped cells with ill-defined cytoplasm and small round to oval hyperchromatic nuclei with small centrally located nucleoli, embedded in a myxoid stroma. The latter is rich in collagen and often contains hemorrhagic foci. A defining special feature is the presence of variably sized vessels that range from small thin-walled capillaries to large vessels Immunohistochemically, most AA express different combinations of estrogen and progesterone receptors, vimentin, desmin, smooth muscle actin CD34, and CD44, but all are negative for S‐100, carcinoembryonic antigen, and keratin.[12].

Wide surgical excision is the treatment of choice .AA is known for local recurrence in approximately 70% of the cases after a period of 2 years postoperatively[13] and it has been reported even 20 years after surgery as well.[6] Han‐Geurts *et al*.[2] proposed the following guideline for treating AA: (1) Complete excision of the lesion when possible, avoiding mutilating surgery, (2) adjunct therapy using arterial embolization and/or hormonal treatment with tamoxifen, raloxifene and gonadotropin-releasing hormone analogues , has been shown to reduce the tumor size needed in case of partial resection of the tumor, and (3) radiation therapy and chemotherapy have limited role due to low mitotic activity however radiotherapy is reserved for cases that are resistant to embolization and/or hormonal therapy and still symptomatic. Due to possibility of late recurrences in future, patients are counselled about the need for long-term follow-up. Magnetic resonance imaging is the preferred method for detecting recurrences.

CONCLUSION

Though AA is a rare entity, it should always be considered, especially when it is an insidious painless vulvar lesion, particularly in premenopausal women in their third to fourth decades of life. High level of suspicion is needed to make a clinical diagnosis. Lowest recurrence rate is achieved if complete resection is possible under any circumstances, which is difficult. AA is rarely life-threatening, and therefore a partial resection when high operative morbidity is anticipated and close and long-term follow-up can result in favourable outcome.

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