Aggressive Angiomyxoma

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ABSTRACT

Myxoid tumors are a heterogeneous group of lesions characterized by a marked abundance of extra cellular mucoid (myxoid) matrix.[1] The term aggressive emphasizes the often infiltrative nature of the tumor and its frequent association with recurrence.[2] A case of aggressive angiomyxoma arising from the vagina in a 55-year-old woman is reported for its rarity.

Key words: Aggressive, angio myxoma, Carney complex

INTRODUCTION

Aggressive angiomyxoma (AAM) is a rare, acquired, benign, neoplasm arising from mesenchymal tissue. The terms "angiomyxoma" and "myxoma" are synonyms.[3] The term aggressive emphasizes the often infiltrative nature of the tumor and its frequent association with recurrence.[2] Under 250 cases of AAM were reported in the literature since the first description of AAM in 1983.[3] A case of AAM in a post-menopausal woman is reported for its uncommon occurrence.

CASE REPORT

A 55-year-old female patient was referred by a gynaecologist with the complaint of a gradually progressive, painless swelling in the vagina for the past 2 years. A mother of four children, she attained menopause 10 years ago and was asymptomatic otherwise. No positive family history of similar complaints was forthcoming.

On examination, there was a glistening, white, gelatinous, non-tender, polypoid swelling, with multiple, superficial ulcerations, protruding out of the vaginal orifice. On close inspection, the tumor was found to be pedunculated, measuring 8 cm long, soft to rubbery in consistency and originating from the anterior vaginal wall [Figure 1a-c]. There was another sessile lesion, 0.5 cm × 0.5 cm, over the posterior vaginal wall. Cervix and uterus were normal. There was no regional lymphadenopathy. The patient, a known hypertensive, on regular treatment, was otherwise asymptomatic. Systemic examination did not reveal any other abnormality. A tentative clinical diagnosis of a malignant tumor was entertained. The tumor was excised and submitted for histopathological examination (HPE).

HPE of the excised mass showed a tumor, composed of fibromyxoid stroma, around proliferating vascular channels of varying size, covered by ulcerated stratified squamous epithelium. Some of the vessels showed marked congestion. The features were suggestive of an AAM [Figure 2a-d].

DISCUSSION

AAM is an uncommon mesenchymal neoplasm, occurring predominantly in the pelvi-perineal region of adults, first described in 1983, by Steeper and Rosai.[4,5] Often reported from the vulvovaginal region, simultaneous abdominal swelling and gluteal region involvement has also been reported.[5] Though 90% of patients are women, involvement in men has also been observed.[6] AAMs are usually solitary, but multi focal involvement has been noticed by various authors,[5,7] similar to our patient. Solitary tumors are reported to be without any systemic abnormalities while multiple lesions could be a manifestation of the Carney complex.[8] The present patient, though having multiple lesions, did not have any clinical evidence of the Carney complex in the form of myxomas at other sites or other endocrine pathology.

The Carney complex, a familial multitumoral syndrome, comprises spotty skin pigmentation, (lentigenes and blue nevi),

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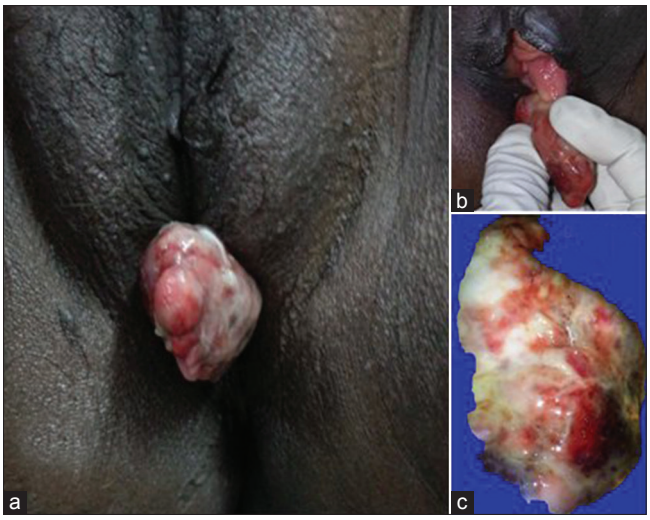

Figure 1: (a and b) A glistening, white, gelatinous, polypoid swelling, with multiple, superficial ulcerations, originating from the anterior vaginal wall, protruding out of the vaginal orifice. (c) An excised, pedunculated mass, about 8 cm long with multiple, superficial ulcerations

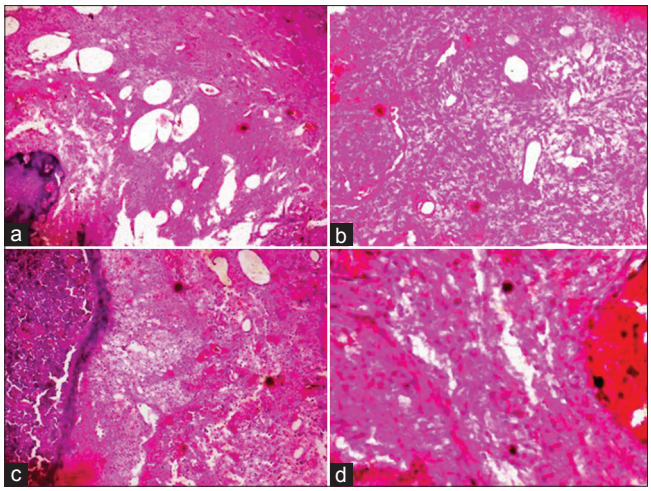

Figure 2: (a) Tumor composed of fibromyxoid stroma around proliferating vascular channels of varying size covered by ulcerated stratified squamous epithelium ×10, H and E. (b) Proliferating vascular channels in fibromyxoid stroma ×10, H and. (c) Surface epithelium covered by Neutrophils ×10, H and E. (d) Congested vascular channel ×20, H and E

myxomas (heart, skin, and breast), endocrine over activity, usually manifested by endocrine tumors, (adrenal cortex, pituitary, testis, and thyroid) schwannomas, and two unusual pigmented tumors, epithelioid blue nevus (skin), and psammomatous melanotic schwannoma (involving skin, viscera or nerve tissue). Carney complex has been linked to chromosome 2p16 and the PRKAR1A gene at 17 q22-24.[9] Cutaneous myxomas range in appearance from small, sessile, opalescent or dark pink papules to large finger-like pedunculated lesions.[1] In the present case, one pedunculated swelling and another sessile lesion were present in the vagina.

Myxomas are not considered as a single entity and eight types are described. AAM is regarded as one of the mainstream soft-tissue myxomas.[10] Angiomyofibroblastoma, cellular angiofibroma, superficial myofibroblastoma and fibroepithelial polyp are other conditions, which occur predominantly over the perineum and may be confused with AAM. The size of the lesion usually >5 cm, hypocellularity, presence of thick walled blood vessels, infiltrative nature, absence of involvement of the superficial epithelium favor the diagnosis of AAM.[10]

The estrogen and progesterone receptor positivity suggests that AAM might be hormone dependent as rapid growth has been observed during pregnancy.[5] The tumor occurs in women in 3rd and 4th decade affecting vulva, perineum, and pelvis.[11] In the present case, patient attained menopause 10 years earlier and the duration of the present illness was only 2 years. It could be presumed that, probably she was unaware of the intravaginal growth until the time it protruded out and became palpable externally.

AAMs must be differentiated from cutaneous focal mucinosis, since, myxomas are true neoplasms, and although benign, may recur after incomplete excision.[8] Metastases are exceedingly rare and overall, the prognosis is good.[2]

Histologically angiomyxoma is a mesenchymal tumor, composed of fibroblasts within a strong myxoid background. Vascular proliferation is also prominent and virtually no mitoses are reported,[12] similar to the findings in the present case. In addition to the characteristic features of AAM, other changes such as ulceration of the squamous epithelium with areas of necrosis observed in our case could be attributed to the infection associated with friction due to chronic exposure. Ulceration of a myxoma has seldom been reported earlier.

The vast majority of cases demonstrate positivity for desmin in the myxoid bundles and/or stromal cells while actions and CD34 may be variably positive.[12] AAMs are locally aggressive and tend to recur in 36-72% after resection.[13] Chromosomal translocation of the 12 q13-15 band involving the High Mobility Group A (HMGA2) gene has been described.[2] The lesions are S100 (nerve) protein negative. Chromosomal analysis and other special histochemical marker studies could not be carried out since the patient could not afford these investigations.

Imaging of these tumors is important to determine the extent and thus the optimal approach to surgery.[5] After the biopsy report was available, the patient was advised Ultra Sonography (USG) and magnetic resonance imaging of the abdomen and pelvis, but the results were not contributory.

Surgical excision as undertaken in our case is the treatment of choice. Gonadotropin releasing hormone agonist therapy seems to have a significant beneficial role, especially as an adjuvant to surgery or where surgery is not possible.[14] Other reported alternative methods of treatment to decrease the

chances of local recurrences are pre-operative angiographic embolization, pre-operative external beam irradiation, and intra-operative electron beam radiotherapy.[15]

AAM has not often been reported in Dermatological Journals. This could probably be because patients with AAM often register with the gynaecologist or a surgeon and rarely, if ever, report to a dermatologist. The diagnosis is often suggested by the pathologist, as with our case.

The case is highlighted for its rarity in dermatological literature.

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