Aggressive Angiomyxoma in African Women: A report of Two Cases

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ABSTRACT
BACKGROUND: Aggressive angiomyxoma (AAM) is a rare soft tissue tumour usually of the perineum. There is no report in the surgical literature of a description of AAM in black Africans.

OBJECTIVE: To report the first description of AAM in black Africa women in the surgical literature and to highlight the value of special immunostains in the complete characterization these rare tumours.

METHODS: Case one was a 38-year-old pre-menopausal woman who presented with a five-year history of a painless mass in the left buttock extending to the left side of the perineum with recent ulceration. Clinical examination revealed a pale and febrile woman with an ulcerated 60 x 40 cm mass distorting the left gluteal region and the left side of the perineum. In case two, a 28-year-old woman reported for the assessment of a pedunculated mass arising from the right labium major that has been present for four years. Clinical examination revealed a 19 x 15.5 cm well-circumscribed mass in the perineum. The mass was completely covered by thickened hairy skin and attached to the right labium major by a short thick stalk that measured 5 cm x 7 cm in size. Both tumours were excised via incisions in the perineum.

RESULTS: In both cases the histopathology of the surgical specimens was reported as bland hypocellular tumours with spindle and stellate cells that lacked mitotic activity consistent with a diagnosis of an aggressive angiomyxoma.

CONCLUSION: The clinical and histopathological features of the tumours described in this report are consistent with a diagnosis of aggressive angiomyxoma. To the best of our knowledge this is the first ever report of AAM in black African women. WAJM 2009; 28(5): 333–336.

Keywords: Aggressive Angiomyxoma, African, Women.

RÉSUMÉ

OBJECTIF: le rapport de la première description de AAM chez les femmes d’Afrique noire dans la littérature chirurgicale et pour souligner la valeur de immunostains spéciale dans la caractérisation complète de ces tumeurs rares.

MÉTHODES: L’une était une affaire de 38 ans avant la vieille femme ménopausée qui s’est présenté avec une durée de cinq ans d’histoire d’une masse indolore dans la fesse gauche s’étendant vers le côté gauche du périnée avec une ulcération des dernières années. L’examen clinique a révélé une femme pâle et fébrile avec un ulcéré 60 x 40 cm de distorsion de masse dans la région fessière gauche et le côté gauche du périnée. Dans deux cas, une femme de 28 ans, signalé pour l’évaluation de la masse pédiculée découlant de la lèvre droite majeur qui a été présent pendant quatre ans. L’examen clinique a révélé un 19 x 15.5 cm masse bien circonscrit dans le périnée. La masse a été complètement recouverte par la peau velue épaisse et attaché à la lèvre droite s’est spécialisé par une tige courte et épaisse qui mesurait 5 cm x 7 cm de taille. Les deux tumeurs ont été excisées par des incisions dans le périnée.

RÉSULTATS: Dans les deux cas, l’histopathologie des pièces opératoires a été rapportée que des tumeurs Bland hypocellulaire à broche avec épaisse couche qui manquait activité mitotique compatibles avec un diagnostic d’une angiomyxoma agressif.


Mots-clés: Angiomyxoma agressif, africaine, des femmes.
INTRODUCTION

Aggressive angiomyxoma (AAM) was first described by Steeper and Rosai some two and a half decades ago (1983). The two authors considered the AAM as a distinct variant of myxoid tumours with a prominent vascular component. This locally invasive and slow-growing tumour has a predilection for the pelvic and perineal regions. Since the original description some 25 years ago, over 65 cases have been reported in the English literature. Most of these reported cases have been in the vuvovaginal region of white females. The patients affected are usually in their reproductive ages. This unusual tumour has a bland histology with few cells that may not have the typical features of malignancy. There is however, high local recurrence rate, therefore, several months of follow up are required after excision in order to detect recurrent tumour. Two cases of this uncommon tumour reported in were seen and treated at the Komfo Anokye Teaching Hospital in Kumasi Ghana. The purpose of the report is to document the first ever cases of AAM in black African women in the English surgical literature, and to highlight the value of special immunostains in the complete characterization of these rare tumours.

CASE REPORTS

Case One

The patient was a 38-year-old premenopausal woman who presented at the outpatient surgical unit with a history of a painless mass in the left buttock for five years. The mass has been growing slowly and over the years has extended to the left side of the perineum. About three months before presentation, she began to experience pains in the mass, especially in the perinael part. Three weeks before presentation she noticed a blister on the mass in the perineum which broke down into a rapidly enlarging and painful ulcer. Clinical examination revealed a pale and febrile patient with a large mass of 60 x 40 cm distorting the left gluteal region in a plexiform shape and extending into the left ischiorectal region fossa of the perineum. An ulcerated and infected area of 10 x 8 cm involving the skin over the left ischiorectal fossa with slough and discharge was noted and gave the impression of a herniating mass. A clinical diagnosis of a soft tissue sarcoma of the left gluteal region with an extension to the perineum and infected ulceration was made. Further investigations included chest X-ray, an ultrasound, an FNAC of the gluteal component and a bacteriological swab for the ulcer. The chest X-ray was normal. The sonogram was reported as a large perineal lipoma. Cytology was inconclusive for malignant cells. Tru-cut biopsy was considered unsafe as the procedure may spread infection in the perineum.

Pathology: The cut surface of the excised specimen was of grey tan, homogenous, and plexiform had a gelatinous appearance (Figure 2). Representative sections were examined by routine H&E stains. The sections were also stained with special immunoperoxidase stain for desmin and smooth muscle actin (SMA) which are biological markers for smooth muscle cells. In addition special staining was done for the detection of CD34 (marker for vascular endothelial cells) and CD117 (marker for gastrointestinal stromal tumours, GIST). Appropriate positive and negative control tissues were included.

H&E sections showed a hypocellular lesion with myxoid stroma, stellate and spindled stromal cells, and scattered variable sized venules and capillaries. No evidence of mitosis or cellular atypia was seen. Immunoperoxidase stains showed 1+ positive for desmin (Figure 3), progesterone, and CD117. The CD34 marker for vascular epithelium was also positive and outlined the blood vessels.

Case Two

The patient was treated with intravenous penicillin G at 24 mega units a day for three days. Transfusion of four units of whole blood was administered. It was considered that excision was the best option to remove the ulcerated and infected tumour. The tumour was excised via a perineal incision that extended from the edges of the large ulcer. It was possible to remove the entire tumour from the perineal and gluteal regions (Figure 2). Intraoperative blood loss was about 400m. The histopathology was reported as AAM.

Figure 1: Clinical presentation of case one with tumour herniating through necrotic perineal skin.

Figure 2: The surgical specimen from case one showing a large plexiform tumour mass with a grey tan and gelatinous appearance.

Figure 3: Desmin, immunoperoxidase. 400x of sections from case one. Positive stain in stromal cell (Arrow).

Figure 4: Clinical presentation of case two. A large pedunculated mass arising from the right labium major.
A 28-year-old woman was referred from the gynaecology clinic to the surgical out-patient on account of a pedunculated mass on her vulva for four years. A diagnosis of an obturator hernia or a lipoma of the vulva was considered. The tumour has been slowly increasing in size since first noticed, was painless and apart from the inconvenience related to the anatomical position, the patient experienced no symptoms. There was no previous history of surgical operations and no previous medical admissions.

Clinical examination revealed a healthy young woman. There was a large polypoid or pedunculated mass 19.0 x 15.5cm arising from the right labium major (Figure 4). The mass had well defined margins, slightly lobulated, firm or rubbery and was attached to the right labium by a stalk measuring 7cm x 5.5cm. The skin over the tumour was thickened. The inguinal lymph nodes were not enlarged.

A chest X-ray was normal. Fine needle aspiration cytology was inconclusive. Due to the anatomical location and the polypoid nature of the tumour no CT scanning was done. The tumour was excised via an incision placed over the right labium. The histopathology was reported as the rare aggressive angiomyxoma with tumour detectable at the excision margins.

**Figure 5**: (Case two). Smooth muscle actin, 400x, immunoperoxidase stain. Positive stain in some stromal cells (see arrow).

**Pathology.** The cut surface of the excised tumour was white, homogenous and had a gelatinous appearance typical of AAM. Representative sections were examined by routine H&E stains as well as immunoperoxidase stains for desmin, smooth muscle actin (SMA), CD34 and CD117. Appropriate positive and negative control tissues were included. H&E sections were similar to those of case one. Immunoperoxidase stains showed 1+ – 2+ positive for SMA (Figure 5).

**DISCUSSION**

Since the first characterization by Steeper and Rosai in 1983 most of the data on the clinical presentation, morphology and histopathology and the diagnosis of AAM has been from case reports and small series. The clinical presentation of the two cases in this report (Figures 1 and 5) was consistent with those from previous reports. In a recent report on soft tissue tumours of the perineum, Behranwala et al indicated the value of imaging studies in the complete evaluation of these often large masses as the physical examination alone may underestimate the extent. On CT scan the AAM shows a well defined margin. It may be distinguished from muscle as the AAM has less attenuation. On MRI, AAM shows high signal intensity on T2-weighted images. These scanning characteristics are related to the high water content of this tumour. The final diagnosis of AAM is based on the features on histopathology. In most published case reports and series the tumours are grossly soft, rubbery and have a gelatinous appearance on the cut section. Microscopically the lesions are sparsely to moderately cellular. The cells have a bland morphology and are mostly stellate, or spindle shaped, embedded in a loose matrix of collagen with scattered vessels of varied caliber. Mitotic activity is absent or infrequent and normal. On immunohistochemical staining AAM cells also show variable expression for vimesin, desmin and SMA and may show ER and PR positivity, but are negative for protein S-100. The macroscopic, microscopic and immunohistochemical features of the tumours described in the two cases in this report (Figures 1–2) were consistent with the features of AAM as described in most previous published reports.

AAM may histologically resemble angiomyoﬁbroblastoma (AMF). AMF is a benign tumour, often smaller than five cm and has well deﬁned circumscribed margins. Microscopically AMF often shows concentric accumulation of stromal cells around the blood vessels. There is considerable similarity in immunostains with both being variably positive for desmin, SMA and CD34. Some authorities believe that both the lesions to be same with a spectrum of biologic aggressiveness.

AAM is a locally aggressive tumour. However, recently cases of fatal metastatic AAM have been reported. AAM is commonly seen in the female perineum and pelvic regions, however it may rarely occur in scrotal areas in males.

Since the AAM is a rare disease there have been no large scale clinical trials to provide evidence based guidelines for the treatment of the disease. Surgical resection is the main treatment modality. The extent of resection is however limited by the need to safeguard and preserve gastro-intestinal and genitourinary functions. Recurrence rates as high as 35–72% have been reported even after resection with clear surgical margins. Further resection for recurrence is possible in many cases. There are reports that some patients with incomplete resections of aggressive angiomyxoma have not developed clinical recurrence after several years of follow-up. The value of extensive surgical resection to obtain tumour negative margins has been questioned. Some reports suggest that postoperative radiotherapy may prevent or delay local recurrence. The best treatment option is complete excision with clear surgical margins whenever feasible.

**REFERENCES**

Aggressive Angiomyxoma in African Women


