Case Report

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Surgery and Clinical Practice

Management of an Exceptionally Giant Aggressive Angiomyxoma in a Developing Country. A Case Report

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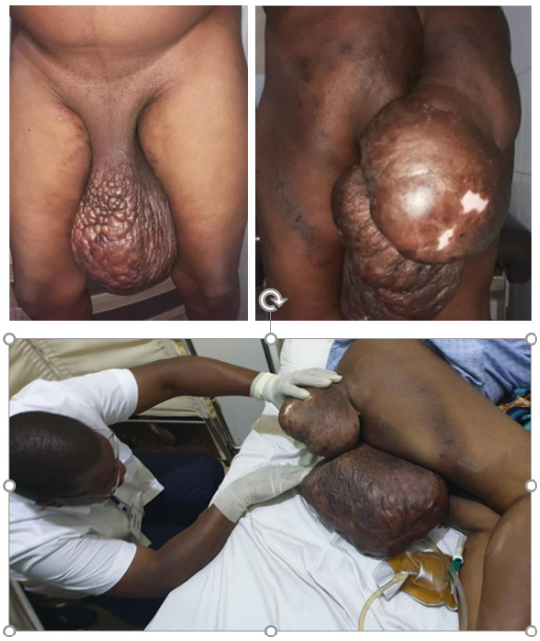
Keywords
Giant angiomyxoma, Female pelvic mass, Developing country, Dyspareunia.

Introduction
First described in 1983, the Aggressive Angiomyxoma (AA) is an uncommon benign soft-tissue tumor usually reported amongst the female population, often occurring from the third decade in the premenopausal period [1]. This slow growing tumor may arise from the pelvis or sub peritoneal area [2] having the capability of reaching wide in size. Symptoms often include compression of the mass on adjacent organs leading to polyuria, pelvic pain and dyspareunia or bowel obstruction [3-5]. The tumor may also progress as a perineal growth [6,7]. One of its main characteristics is the high local recurrence despite extensive surgery. We report a case of an exceptional recurrent giant AA in a woman living in a developing country.

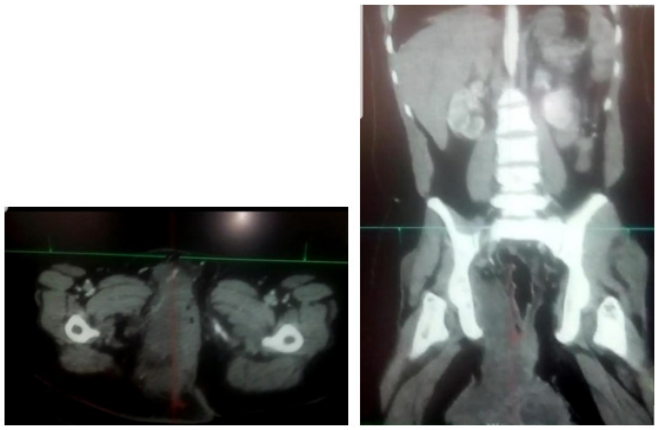
Clinical Presentation
A 43-year-old patient with no previous medical history presented a perineal mass evolving for nearly 10 years. It was a third recurrence after multiple surgical procedures. This mass was bilobed, protrusive and solid, located in the right perineal region with an anterior component leading to a downward pulling of the vagina. The patient had no urological, digestive or gynaecological disorders.

Blood tests showed a haemoglobin rate of 8 g /dl and no other disorders. The abdominal and pelvic CT scan revealed a vascularized fat-toned mass extending spindle-shaped into the ill-defined ischiorectal fossa with no secondary lesion at a visual distance.



Diagnosis and Treatment

Under general anesthesia, we proceeded to resection the bilobed mass via the perineal route and a loose closure with diverting colostomy. The patient was transfused of 2 units of red blood cells during the operation. The bilobed mass weighed 8.8 kilograms. Histology results revealed an "aggressive angiomyxoma".



Outcome

The postoperative follow-up was simple. The patient resumed normal bowel function on day 3 and completed perineal healing within two months. After the seventh month post resection, we closed the stoma and repaired an umbilical incisional hernia. Once again, the operative follow-up was simple with the patient regaining bowel function on day 4 and healing of the operative wounds on day 10.

Discussion

AA is a benign myxoid infiltrative tumor, which often concerns middle aged women [1]. However, AA has been reported in preadolescent [8] and menopausal women [9]. This tumor usually affects the pelvic organ, the sub peritoneal area or the perineum [3]; however, rare ectopic areas such as the liver [10] or maxillary glands [11] have also been described as well as a few cases of AA amongst the male population involving the scrotum or prostate [12]. The abdominal and pelvic CT scan is the main radiologic exam which can assess the depth and the outline of the tumor. A magnetic resonance imaging (MRI) may help with the differential diagnosis highlighting hyperintense T2 signal intensity [13]. Metastases are uncommon but had been reported [14,15].

A histopathological examination showed a myxoid stroma made up of numerous blood vessels with thick walls and fibroblasts involving adipose tissue surrounding it [16]. So, the tumor is not well defined and complete resection is not guaranteed, in turn leading to a high risk of recurrence. There is no evidence of cytologic atypia, no evidence of mitosis or necrosis which confirms the benign nature of the tumor [4]. The main treatment of AA is surgery. Alternatives treatment such as radiotherapy for extensive deep lesions or as an adjuvant treatment to limit recurrence had been supported in previous papers [17]. Other authors used the gonadotropin-releasing hormone agonist as a neoadjuvant treatment or to prevent recurrence in some type of AA tumors [18]. Chemotherapy demonstrated no efficiency for this tumor without mitotic implication [19]. The morbidity of AA is essentially depending on the site surrounding impact with a high risk of reoccurrence up to 50% [20]; mostly within the 3 years after surgery.

We have probably described one of the most giant AA reported in the literature with a bilobar component reaching a total weight of almost 9 kilograms. This impressive size was favored by the precarious environment, which delayed the management of this tumor and limited radiological investigation. After several previous superficial surgeries, the patient lived 10 years with recurrent tumors before the providential surgery. This involuntary long-time evolution confirmed the benign nature of the lesion. However, the impressive size really impaired the quality of life of the patient leading to a near social death. We chose to protect the perineal wound with a temporary lateral colostomy to optimize healing in the context of remote areas with limited healthcare. The adjuvants treatments to limit recurrence are unfortunately less available in emerging countries. The best approach for the patient in this case remains a clinical and radiological follow-up with iterative surgery to prevent the tumor from reaching a significant size.

Conclusion

The AA is a benign tumor which can reach an exceptionally large size and impair quality of life. Surgery remains the gold-standard treatment. Nevertheless, some other neoadjuvant or adjuvant therapies can improve the global management. Despite a large resection, the risk of local recurrence remains high. For emerging

countries with low health infrastructure, a clinical and radiologic monitoring approach with early iterative resection could be a good compromise.

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