Aggressive rhabdomyosarcoma of the vulva in a young Sudanese woman

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ABSTRACT

A 22-year-old para 2, female presented with a 2-month history of a progressively vulvar mass. Clinically, her general condition was poor. She had bilateral inguinal lymphadenopathy. Local examination revealed a large deeply infiltrating vulvar mass. Pathological evaluation revealed pleomorphic rhabdomyosarcoma.

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Sarcomas arising in the vulva are rare comprising between 1.5% and 5% of all vulvar malignancies.[1] Leiomyosarcoma is the most common histological variant of these tumors.[2] Since the disease was rare; the literature revealed only a few cases. Much of the knowledge comes from sporadic case reports and a small number of reviews. Usually they present as painless vulvar swelling and diagnosed by biopsy and histopathology. We are reporting a lethal case of a huge, rapidly growing vulvar rhabdomyosarcoma.

Case Report. A 22-year-old para 2 Sudanese lady was referred to Khartoum hospital with a very huge rapidly growing vulvar swelling of 2 months duration. She looked ill, wasted, and pale. Local examination revealed a huge nodular firm vulvar swelling with necrotic ulcers on its wall. It measured 17 cm x 23 cm x 21 cm (**Figure 1**). Anatomically, it had been related to the site of left labium minus. The inguinal lymph nodes were enlarged on both sides, more so on the left side which showed an ulcer on the surface. There were dilated veins draining up from the pelvis to the thorax. Biopsy from the vulvar mass had been taken. Macroscopic examination showed a lobulated mass of soft tissue measuring 4 x 3 x 2 cm. The specimen was fixed in 10% formalin and blocks were processed by paraffin wax method, sections were stained by hematoxylin and eosin. Histopathological and immunohistochemical examinations revealed "an ulcerative mass consisting of spindle shaped and round cells in a myxomatous stroma. The cells nuclei are large and they show mitotic activity. Some cells show plentiful acidophilic cytoplasm with cross striations. There were scattered mitosis. Pleomorphic and immunoreactivity (positivity) for polyclonal desmin and muscle specific actin (MSA), negative for vimentin leucocytes common antigen (LCA), CD99 and neuron specific enolase (NSE).

Discussion. Mesenchymal tumors of the vulva both benign and malignant are rare. Behranwala et al[2] reported 17 cases of vulvar tumors in a 16 year period. Leiomyosarcoma was the most frequent type (5 cases) and only one case of rhabdomyosarcoma was reported. The natural history of vulvar sarcomas is not clearly understood. This is related to the rarity of the vulvar involvement by primary sarcomatous neoplasms, various histological types and the limited number of the reported cases. In addition, is it difficult to draw a conclusion as most of the reported cases had a short term follow up. In a previous report,[2] the patient's age was 22 years and the mean age was 40 years. Metastatic lesions in patients with a primary carcinoma of the vulva are generally rare, but osseous metastases arising from a vulvar carcinoma have only been reported in a small number of cases in the literature.[3] The patient died a few days after admission (even before completing her investigations and grading of the tumor), which indicated the aggressive of

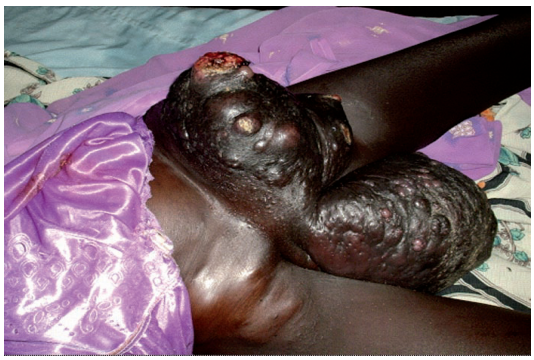

Figure 1 - Showing a large ulcerated tumor of the vulva.

the tumor. However, low-grade sarcoma of the vulva was reported before.[4] After a proper counselling the patient was screened for HIV and was negative. Due to HIV epidemic in African countries, sarcomas had changed their pattern and presentations and HIV infection was associated with significantly increased risk of cancers of the vulva.[5]

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