

Massive primary leiomyosarcoma of the testis

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Abstract:

Primary leiomyosarcoma of the testis is a very rare tumor with only 9 reported cases in the English literature, of which none has been reported in patient of African descent. In this report we described the first case of a massive primary leiomyosarcoma of the testis. A 20-year-old-man presented with huge fungating tumor of the left scrotum with no clinical or imaging evidence of regional or distance metastasis. Left radical orchiectomy was performed, followed by satisfactory postoperative recovery. The postoperative histopathology examination and Immunohistochemistry for desmin, actin, vimentin, and S-100 indicate primary leiomyosarcoma of the left testis. Eleven month later the patient developed convulsion episodes. Brain CT scan and MRI showed brain metastatic deposition. Abdominal ultrasound indicates involvement of paraaortic lymph nodes. The patient received 5 cycles of vincristine, actinomycin D, and cyclophosphamide in addition to Epanutin 50 mg three times a day. However, Twenty two month after surgery the patient underwent another surgery for local recurrence. The present case showed aggressive course of primary testicular leiomyosarcoma in contrast to the benign course previously reported. In addition to radical orchiectomy with high ligation of the spermatic cord, systemic chemotherapy, and regional radiotherapy may be the treatment of choice for some primary leiomyosarcoma of the testis.

Introduction

Although leiomyosarcoma can arise from any tissues that contains smooth muscle. primary leiomyosarcoma of the testis is extremely rare tumor compared with the paratesticular leiomyosarcoma that originate from spermatic cord, epididymis, or tunica vaginalis [1] Worldwide, only 9 primary testicular leiomyosarcoma cases have been reported in the English literature [2-10]. According to the very limited cases, the usual age of occurrence is more than 50 years, the tumor involve one side in most cases, and can adequately be treated by radical inguinal orchiectomy preceded by high ligation of the spermatic cord [11]. In most reported cases including the massive ones, the disease was localized with no regional or distance metastasis, and the prognosis seems to be good if radical resection is performed. In this report we present the 10th case of primary testicular leiomyosarcoma with aggressive clinical course, indicating the need for aggressive multitherapy approach in some primary testicular leiomyosarcoma.

Case presentation

A 20-year-old-man admitted to the hospital complaining of massive and fungating tumor of his left scrotal (Figures 1). The patient has been refer from a regional hospital, where he has been admitted for nearly one month before referring him for further investigation and management. The history of his present illness started 2-year-early with small painless lump in his left scrotum that increased gradually in size. Physical examination revealed huge infected and fungating tumor measuring 20x30 cm on the left testis (Figures 1), no superficial lymph node swelling or gynecomastia were noted. Liver function and tumor markers, including alpha fetoprotein, lactate dehydrogenase, and human chorionic gonadotrophin assays, were all within normal ranges. Abdominal ultrasound and chest X-ray showed no evidence of local, regional or distant malignant lymphadenopathy or retroperitoneal metastasis. High ligation of the spermatic cord and left radical orchiectomy and excision of the tumor en bloc with the involved skin was performed using inguinoscrotal approach. Macroscopic examination of the tumor revealed solid fleshy massive tumor measuring 20x30 cm, yellowish-white in color, (Figures 1), with areas of hemorrhage and necrosis noted. The histology examination showed consisted of monomorphic pattern of interlacing bundles and fascicles of spindle cells. The cells contained oval and cigar-shaped vesicular nuclei, with inconspicuous nucleoli (Figure 2). Atypical cells were not prominent but occasional cells showed multinucleation. Mitotic figures were present over 20 per 10 high power fields. Subsequent Immunohistochemistry staining of the tumor was positive for actin, desmin, and vimentin, but not for S-100 (Figure 3-7). The patient had an

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uneventful postoperative course and received no adjuvant therapy. However 11 months later the patient was readmitted following 2 episodes of epileptic attacks associated with loss of consciousness, and left hypochondria swelling. Abdominal ultrasound revealed distant metastasis involving the paraaortic lymph node. The brain CT and MRI scan showed metastatic deposition

(Figure 6). Abdominal ultrasound indicates involvement of paraaortic lymph nodes. The patient received and responds well to five cycles of chemotherapy (vincristine, actinomycin D, and cyclophosphamide), and Epanutin tablets (50 mg) three times a day. Twelve months later the patient developed local recurrent that treated with excisional surgery followed by local radiotherapy

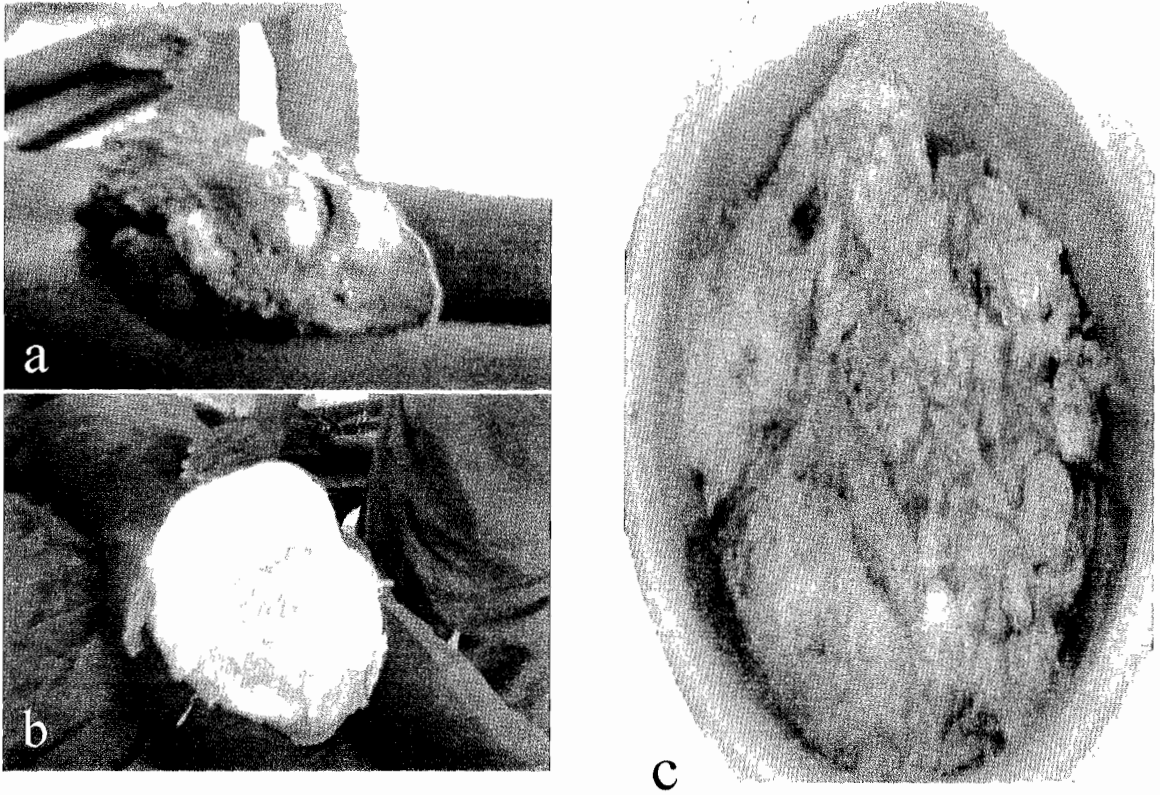


Figure 1. (a) Massive fungating 20x30 Lt. scrotal tumor (b) The tumor rapped with white gauze (c) The tumor after the surgery showing yellowish-white color and areas of hemorrhage and necrosis

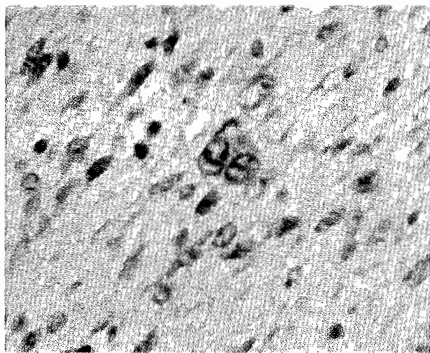


Figure 2. H & E spindle shaped cells with cigar-shaped nuclei with bi-nucleation of the cell at the middle Mitotic figures seen (X40).

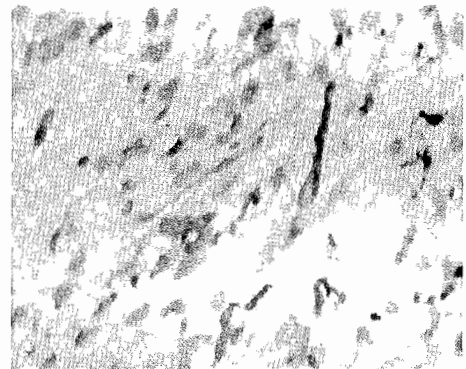


Figure 3. Immunohistochemical stain for desmin positive in spindle cells (X40)

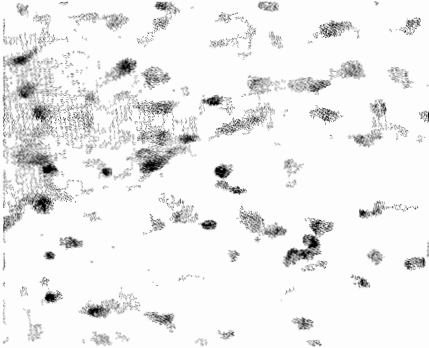


Figure 4. Immunohistochemical staining for smooth muscle actin positivity in spindle cells (X40)

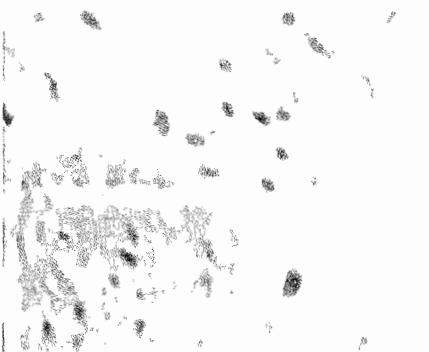


Figure 5. Invasion of the dartos muscle X40

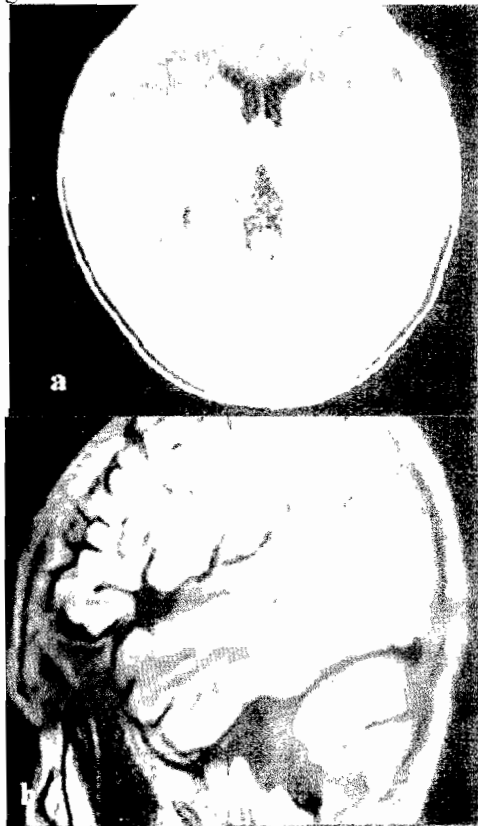


Figure 6. (a) CT axial image with contrast (b) Saggital MRI Flair image showing Rt. occipital cortical lesion with perilesional edema

Discussion

Leiomyosarcoma is extremely rare tumor of the testis. Although more than 100 paratesticular leiomyosarcoma have been reported in the literature [1] with 80% arising from the soft tissue of the spermatic cord and 20% originating from the epididymis or darts of the scrotum, few cases of primary leiomyosarcoma of the testis have been reported. To our knowledge, only 9 cases of intratesticular leiomyosarcoma have been reported in the English literature [5;12-19] . However, all were reported in Caucasian thus making the present case unique in sense that it is the ever first primary intratesticular leiomyosarcoma in African black race.

The etiology behind primary testicular leiomyosarcoma is not yet clear, however according to the previous limited literature, high-doses of anabolic steroids and chronic inflammation are the main risk factors for intratesticular leiomyosarcoma [20;21]. Anyhow, our patient denies any past history of chronic inflammation or long interval steroids therapy.

Most reported testicular leiomyosarcoma had early clinical stage (stage 1). The macroscopic feature, as in our case, revealed solid gray-white tumors. The reason behind early clinical stage could be the exposed anatomic position that allows easy recognition and the slow-growing feature of testicular leiomyosarcoma. However, in some cases the disease might spread via local invasion, lymphatic dissemination, and/or hematogenous metastasis. Regional lymph nodes, including retroperitoneal nodes, are the most commonly affected.

In most reported cases radical orchiectomy is the standard treatment and usually no adjuvant therapy was needed. The two exceptions were additional retroperitoneal lymph node dissection in one case, and chemotherapy in another case (Hachi et al., 2002). Metastasis was another exception in reported cases and only one case developed pulmonary metastasis 14 months after surgery. The present case exhibits an aggressive natural history with propensity for local recurrent and distant metastases. Although, standard therapy for intratesticular leiomyosarcoma is difficult to recommend due to the few reported cases, in some cases like the present one aggressive and multitherapy approach may be needed

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