Adult paratesticular malignant fibrous histiocytoma treated with surgery, systemic chemotherapy and postoperative adjuvant radiotherapy

ABSTRACT
Paratesticular malignant fibrous histiocytoma is an extremely rare malignancy of the scrotum. This malignancy has rarely been described in the literature and scant information exists on the optimal management of this cancer. We present here a case of a 57-year-old man with a diagnosis of high-grade malignant fibrous histiocytoma of the left intrascrotal region who underwent radical orchietomy, systemic chemotherapy and postoperative radiotherapy.

KEY WORDS: Fibrous histiocytoma, intrascrotal, malignant, paratesticular, radiotherapy

INTRODUCTION
Paratesticular malignant fibrous histiocytoma is an extremely rare malignancy of the scrotum. This malignancy has rarely been described in the literature and scant information exists on the optimal management of this cancer. We present here a case of a 57-year-old man with a diagnosis of high-grade malignant fibrous histiocytoma of the left intrascrotal region who underwent radical orchietomy, systemic chemotherapy and postoperative radiotherapy.

CASE REPORT
A 57-year-old gentleman who presented with a diagnosis of high-grade paratesticular malignant fibrous histiocytoma. He originally presented with a left descended testicle. In 1979, he underwent a left orchiectomy. He had an uneventful genitourinary history until 1996 when he presented with a left intrascrotal mass. This mass was gross totally excised and pathology revealed benign lipoma.

He was without evidence of disease until February of 2005. At that time, the patient noted a left-sided intrascrotal mass. His primary care physician confirmed this finding and an ultrasound revealed a 1.4 cm solid lesion in the left scrotum. In June of 2005, the patient presented for resection of this lesion. Pathology showed a 5.1 x 4.9 x 3.6 cm mass consistent with high-grade malignant fibrous histiocytoma. Margins of excision were negative. The patient then underwent a course of systemic adriamycin-based chemotherapy. Further workup via ultrasound revealed a 1.1 cm hypoechoic lesion, which was contiguous with the right testicle. The patient then underwent a right radical orchiectomy in November of 2005 with re-excision of his prior surgical scar. Pathology was entirely negative for residual sarcoma. The patient then presented for a course of postoperative radiotherapy.

The patient was treated with a course of external-beam radiotherapy to the pelvis delivered in an AP/PA fashion with the inguinal lymph nodes inclusive. 6 MV photons were delivered from the anterior field and 15 MV photons were delivered from the posterior field. The radiation dose was calculated from the posterior exit beam to delineate dose delivered to the inguinal nodes. Then, customized electron cutouts were designed to bring the dose to the inguinal nodal chain to 100% of the prescribed dose. 9 MeV electrons were utilized. Depth of the inguinal nodes was calculated from the treatment planning CT scan.

By 1440 cGy, the patient developed some mild groin redness. By 2520 cGy, he developed groin erythema and discomfort and was given Tylenol #3 in addition to topical ointments. At 3060 cGy, the patient developed a tinea infection of the groin and was subsequently placed on an anti-fungal regimen. At
this juncture, the patient also required a significant radiation therapy break, secondary to moist groin desquamation and discomfort. In addition, penile erythema and discomfort were noted. The patient was prescribed a course of Silvadine cream to be applied to the affected areas while on break. Upon return to clinic, the patient experienced near total re-epithelialization and resolution of his groin and penile erythema. He completed a dose of 4500 cGy without further difficulty. However, the patient elected to forgo any subsequent boost and treatment was discontinued at 4500 cGy. At one month, follow-up, the patient demonstrated no evidence of malignancy and near-full resolution of treatment side effects. Additionally, axial computed tomography scans of the abdomen and pelvis, respectively, showing absence of malignant adenopathy at six weeks follow-up.

**DISCUSSION**

This case critically underscores the difficulty in managing malignancies requiring pelvic and inguinal nodal radiotherapy and their attendant side-effects. Paratesticular sarcomas are an extremely rare entity and a paucity of information about their management exists in the literature. Fagundes, et al analyzed 18 patients with this disease entity from the Massachusetts General Hospital. 16 patients underwent radical orchietomy, while all 18 underwent surgical resection. Nine of the 18 patients received adjuvant radiotherapy coupled with orchietomy, while the remaining nine patients received surgery alone. Five of the nine patients treated with orchietomy alone developed locoregional recurrence, while no patient who received adjuvant radiotherapy relapsed. The median radiation dose was 4500 cGy.

In a review from the Princess Margaret Hospital, 21 patients with adult paratesticular sarcoma were analyzed. 14 patients presented with primary disease. 13 of these 14 patients underwent initial radical orchietomy. Six of these 14 patients underwent either adjuvant surgery or adjuvant radiotherapy to the groin or to the groin and scrotum and none of these six had local relapse. RT doses ranged from 2400 cGy to 6000 cGy. Of the seven patients referred with recurrent local disease only one could be salvaged.

In another case report by Lissmer and colleagues, a patient underwent wide- excision orchiopexy to the inguinal region and radiotherapy for a scrotal liposarcoma. The patient’s radiotherapy had to be discontinued at 3800 cGy secondary to abscess formation and decubitus around the silicone wrapped testicle. The patient was followed for 30 months following surgery with no evidence of tumor recurrence.

These three studies illustrate the importance of adjuvant therapy to the management and prevention of recurrence of intrascrotal sarcoma. Furthermore, the Princess Margaret Hospital review demonstrates the need for adjuvant therapy to the groin to prevent relapse in these patients.

Additionally, Froehner and colleagues reviewed 22 cases of locally recurrent malignant fibrous histiocytoma and found the prognosis for these patients to be extraordinarily poor. Only two of 22 patients survived past 3.5 years of follow-up. This study demonstrates the need for proper multi-modality treatment of patients presenting with primary malignant fibrous histiocytoma so as to avoid the morbidity that accompanies local recurrence.

This case of scrotal malignant fibrous histiocytoma illustrates the critical importance of adequate primary and adjuvant management as well as proper radiation therapy technique. Also, critically important is the meticulous care of the patient during radiotherapy as adjuvant radiation therapy is often not without significant side effects in this anatomic site. Intensity modulated radiation therapy, as well as conformal therapies, may play a role in managing this difficult entity in the future. Further retrospective reviews and, possibly, prospective trials are warranted to more fully elucidate the optimal management of this rare malignancy.

**REFERENCES**


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