Paratesticular myxoid liposarcoma in a 23-year old Nigerian

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Abstract

Paratesticular liposarcomas are rare tumors and are usually seen in patients in middle age or older. Optimal treatment is radical orchidectomy. Radiotherapy or chemotherapy is added for advanced disease or recurrences. These practice guidelines often vary from the experience in developing countries.

We present a 23-year old man who presented with paratesticular myxoid liposarcoma, after transscrotal orchidectomy for “testicular tumor” without histology. He was subsequently managed by neoadjuvant chemotherapy and complete tumor excision.

A case of paratesticular myxoid liposarcoma in a young man is highlighted. Also noted is the fact that complete extirpation at primary surgery reduces the risk of local recurrence. The practice of transscrotal orchidectomy and non-submission of surgical specimens is highlighted and condemned.

Introduction

Paratesticular myxoid liposarcomas are rare tumors that occur in middle aged men. These tumors are prone to local recurrence following inadequate extirpation, even with adjuvant radiotherapy. Mean sizes ranging from 5 cm to 11.7 cm have been reported.1,2 There is agreement that the management should consist of radical orchidectomy with adjuvant radiotherapy with or without chemotherapy.

We report a case of paratesticular myxoid liposarcoma in a 23-year old man who had a transscrotal excision of a “testicular tumor” seven months earlier, with no histopathology of the specimen. He was managed with neo-adjuvant chemotherapy, complete tumor excision and hemiscrotectomy. We believe this case is unique because of the patient’s age, which is a departure from the norm. We also wish to highlight the harmful practice of transscrotal orchidectomy for testicular neoplasms and associated local recurrence that occurs in the developing world.1 The problem of non-submission of surgical specimens is also highlighted.

Case Report

A 23-year old man presented with a 4-month history of left hemiscrotal swelling. He had had a transscrotal orchidectomy seven months previously for a “testicular tumor” in a rural clinic. The specimen had been discarded.

Examination showed a firm, globular, left intrascrotal mass (14x11x6 cm) with attachment to skin, ulceration and enlarged left inguinal lymph nodes (Figure 1). Ultrasonography suggested a malignant testicular tumor with inguinal and retroperitoneal lymph node metastasis. Computerized tomography was not performed because the patient could not afford the $450 fee. Other investigations, including alfafetoprotein and beta human chorionic gonadotropin assay, human immunodeficiency virus screening, full blood count and chest X-ray were all normal.

An impression of locally recurrent germ cell tumor of the testes was made and the patient started on a neo adjuvant regimen of bleomycin, etoposide and cisplatin. Three courses were given with marked regression in size of the inguinal lymph nodes, softening of the scrotal mass and restoration of the overlying skin mobility, with necrosis and ulceration (Figure 2). Complete tumor excision, inguinal node dissection and hemiscrotectomy was subsequently performed. On surgery, about 50% of the tumor was necrotic (Figure 3). The specimen measured 20x14x6 cm and weighed 856 grammes. Primary skin closure with drainage was made (Figure 4). Post-operative recovery was good.

Histology shows a malignant lipomatous proliferation of cells with a small vesicular nucleus. Characteristically, many of these cells are lipidplasts which contain lipid vacuoles in their cytoplasm which indent the nucleus or displace it to an eccentric position (see histology in Figure 5). The stroma shows a characteristic plexiform arrangement of capillaries in the stroma as well as abundant myxomatous matrix (see histology in Figure 6). Fibrous tracts divide the tumor into lobules and there are extensive areas of coagulative necrosis.

The tumor has a pushing border. The patient was counseled on plans for further chemotherapy, and radiotherapy. He, however, requested to be discharged because he did not have enough money to continue treatment. He has since been lost to follow-up.

We did not perform contralateral inguinal node dissection as there was no clinical evidence of their involvement, and they may be contaminated with tumor if this is performed at the time of tumor resection.

Retroperitoneal lymph node dissection was not offered immediately because we wanted to control local disease and ascertain tumor histology before selecting adjuvant treatment.

Figure 1. Tumor at presentation.

Figure 2. Tumor after chemotherapy. Note the absence of lymph nodes.
Discussion

Paratesticular liposarcomas are rare tumors and are often reported as isolated cases. They refer to liposarcomas arising from the spermatic cord, testicular tunics and epididymes, and usually occur in men aged 41 years to 87 years. Tumor size varies from 3 cm to 30 cm (mean 11.7 cm). These facts are at variance with the findings in our patient who was 23 years old at presentation. The reasons for this are unclear. Its occurrence should cause doctors to consider it as a differential diagnosis of testicular cancer in young men. This is important because we offered our patient neoadjuvant chemotherapy based on a presumptive diagnosis of testicular germ cell tumor. Since complete surgical resection offers the best chance of cure for these patients, and the established method for orchidectomy for testes/cord cancer is through an inguinal incision, the practice of orchidectomy via transcrotal incision by unqualified personnel as occurred in our patient is to be condemned. This procedure leads to tumor spillage and contamination of scrotal skin lymphatics with subsequent enlargement of inguinal lymph nodes and significant morbidity.

The fact that the initial specimen had not undergone histological examination is an unfortunate practice that compromises management. We gave neoadjuvant chemotherapy to reduce tumor bulk, improve resectability and proceeded to surgical excision later.

Since paratesticular site and tumor size less than 5 cm is a good prognostic factor for sarcoma survival, our patient’s prognosis would have been better if he had been offered optimal care at the initial surgery. It is, therefore, important to emphasize mandatory continuing medical education for doctors so as to minimize these occurrences. Paratesticular myxoid liposarcoma is a rare neoplasm that can occur in young men and the best chance of cure is by radical orchidectomy. Histopathological examination of the specimen is mandatory to define tumor type and plan further treatment. When local recurrence has occurred, neoadjuvant chemotherapy with complete tumor excision is a useful method of achieving local control.

In conclusion, paratesticular liposarcoma is a rare neoplasm that can occur in young men and the best chance of cure is by radical orchidectomy. Histopathological examination of the specimen is mandatory.

References

1. Montgomery E, Fischer C. Paratesticular liposarcoma: a clinicopathologic study. Am J Surg Pathol 2003;27:20-47.
2. Ozkara H, Ozkan B, Alici B, et al. Recurrent paratesticular liposarcoma in a young man. Urology 2004; 171:343.
3. Mondaini N. Palli D, Saieva C, et al. Clinical characteristics and overall survival in adult genitourinary sarcomas treated with a curative intent: a multicenter study. Eur Urology 2004;47:468-73.
4. Ugwumba FO, Nnabugwu I, Nnakenyi EF, et al. Giant Seminoma following Ipsilateral Orchidectomy: a rare finding. Case report. Eur J of Cancer Care 2009;18:322-4.
5. Rowland RG, Herman JR. Tumors and infectious diseases of the testes. In: Gillenwater JY, Grayhack JT, Howard SS, Mitchell ME, editors. Adult and pediatric urology, 4th ed. Philadelphia: Lippincott Williams and Wilkins; 2002.