Diffuse large B cell lymphoma of the spermatic cord: a case report and literature review

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Primary lymphoma of the spermatic cord is rare. We report a case of primary lymphoma of the spermatic cord and review the literature. A 77-year-old man presented with a 5 month history of an enlarging right inguino-scrotal mass. On physical exam, the mass involved the spermatic cord. Computerized tomography and magnetic resonance imaging revealed a 10 cm x 4 cm inguinoscrotal mass related to the spermatic cord. A right inguinal orchidectomy with wide local excision was performed. Pathological and immunohistochemical evaluation identified the tumor as a diffuse large B cell lymphoma of the spermatic cord. Postoperatively, patient began a chemoradiotherapy regimen consistent with metastatic lymphoma.

Key Words: spermatic cord, lymphoma

Introduction

Tumors of the spermatic cord are uncommon, and primary lymphoma of the spermatic cord is rare. In the international English medical literature, only 16 cases of primary lymphoma of the spermatic cord have been reported.[1] We provide an additional case of primary lymphoma of the spermatic cord.

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Case report

A 77-year-old man presented with a right inguino-scrotal mass. The patient noticed a right scrotal mass 5 months earlier and it has been increasing in size. On physical examination, the patient had a painless palpable hard mass in the right inguinoscrotal region involving the right spermatic cord and extending to the right proximal corpora. Computerized tomography of the abdomen and pelvis revealed a 10 cm x 4 cm inguinoscrotal mass involving the spermatic cord. No enlarged lymph nodes or distant metastases were noted, Figure 1. Magnetic resonance imaging (MRI) showed a right inguinal scrotal mass, which could

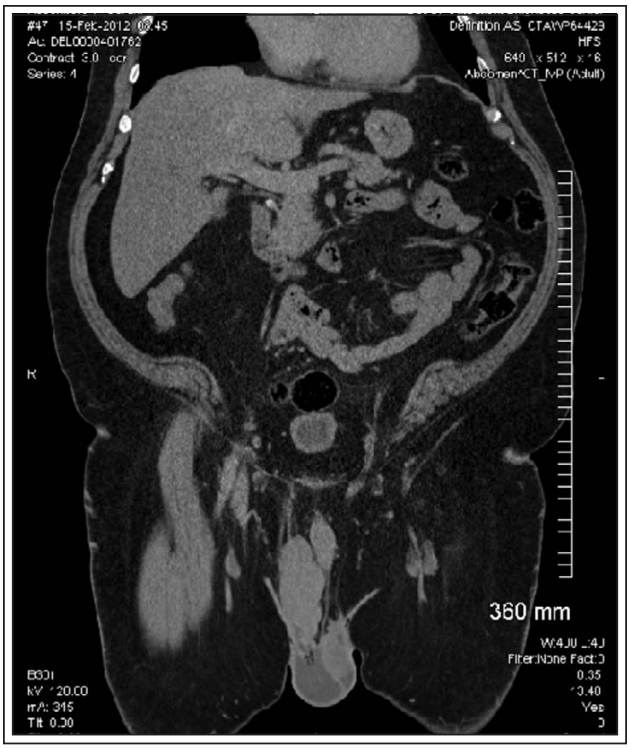

Figure 1. Preoperative computed tomography scan showing the large inguino-scrotal mass.

not be differentiated from the testicle, Figure 2. Given the presentation and age of the patient liposarcoma

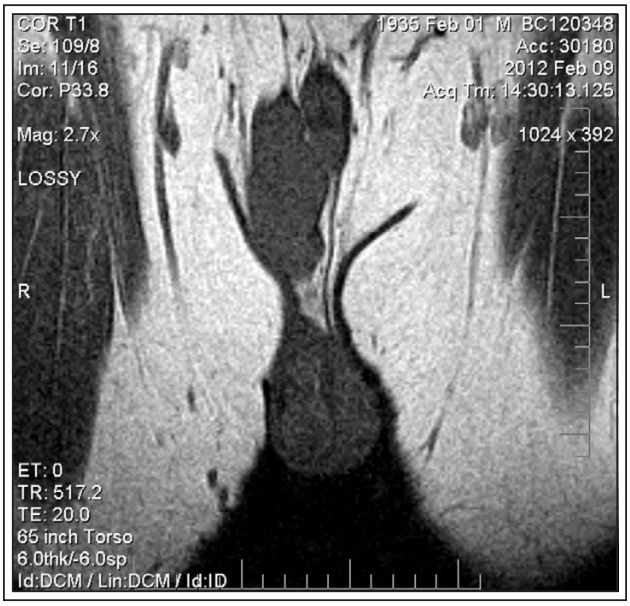

Figure 2. MRI imaging showing a 10 cm x 4 cm inguino-scrotal mass without lymphadenopathy.

of the cord was suspected. The patient underwent a right inguinal orchiectomy with wide local excision. The mass was confined to the spermatic cord and did not involve the testis. Frozen section suggested the diagnosis of lymphoma. Surgical margins were negative. The tumor measured 10 cm x 8 cm x 4 cm and was whitish-yellow and firm, Figure 3.

Histopathological examination revealed diffuse large B cell lymphoma non-germinal center immunophenotype. The tumor was positivite for CD20a, CD79a, BCL-6, BCL-2, MYC, and MUM1 tumor markers. The ki-67 proliferation labeling index was estimated to be greater than 80%. Immunostaining was negative for CD3, CD5, CD10, and CD30. The margins of the spermatic cord were negative.

Given the diagnosis of primary lymphoma, the patient underwent further postoperative staging. A bone marrow aspirate and biopsy were negative. MRI demonstrated a benign cyst in the posterior cranial fossa. Positron emission tomography - computed tomography (PET-CT) showed high grade multifocal hypermetabolic tumor activity involving the left adrenal gland and left para-aortic lymph nodes suspicious for metastasis. The patient was started on rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP).

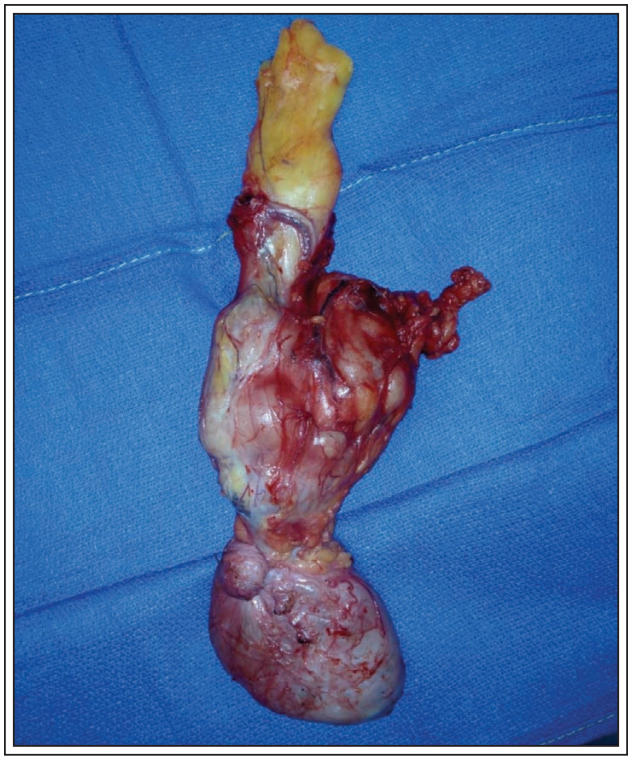

Figure 3. Gross photo of radical orchiectomy specimen.

Discussion

Male gonadal non-Hodgkin's lymphoma is relatively uncommon and is estimated to account for less than 2% of all lymphomas.[2] Primary lymphoma of the spermatic cord is extremely rare, with only 16 cases reported. This rare cause of inguinal or inguinoscrotal mass is commonly misdiagnosed as an inguinal hernia.[3] Patients with lymphoma of the spermatic cord typically present with a palpable mass in the inguinal canal or scrotum.[4] Generalized symptoms typical of lymphoma include weight loss, night sweats, and fever and are not typically seen in these cases.[5] Eighty percent of primary lymphomas of the spermatic cord are non-Hodgkin DLBCL and have a poor prognosis.[3]

Given the rarity of primary spermatic cord lymphoma and the similarity to primary testicular lymphoma (PTL), treatment is based on treatment for PTL. The initial treatment is high inguinal orchiectomy. Survival is improved with the addition of systemic chemotherapy, especially anthracyclines.[6] For decades, the primary therapy has been cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP) regimen. Recently, the addition of rituximab, a chimeric anti-CD20 monoclonal antibody, to the CHOP regimen (R-CHOP) has shown marked improvement in progression-free and overall survival.[7] CNS relapses in both the brain and meninges are common with a 5 and 10 year risk of 19% and 34%, respectively, for patients with PTL.[8] Consequently, intrathecal prophylaxis is recommended as it has been shown to reduce central nervous system relapse. It is typically administered as high dose intrathecal methotrexate (IT-MTX) for improved penetration of the brain parenchyma.[9] Prophylactic radiotherapy of the contralateral hemiscrotum is recommended as it may reduce the incidence of relapse at this site.[2,8] It has been recently shown that combination therapy with R-CHOP, IT-MTX, and prophylactic radiotherapy yields a 5 year progression free and overall survival as high as 74% and 85%, respectively, for patients with stage I or stage II PTL.

For patients with advanced PTL, treatment is R-CHOP chemotherapy, prophylactic scrotal radiotherapy, and intrathecal chemotherapy. Since our patient likely has advanced primary lymphoma of the spermatic cord, he is currently undergoing combination chemoradiotherapy based on therapies for advanced PTL. A review of previous cases has indicated a poor prognosis, however treatment has been limited to orchiectomy alone or, in some cases, combined with chemotherapy.[3,9,10] Chemotherapy including radiotherapy with CNS prophylaxis may improve outcomes in primary lymphoma of the spermatic cord. □

References

1. Diakatou E, Haramis G, Kostopoulou A, Kakiopoulos G. Primary lymphoma of the spermatic cord: A case report and review of the literature. *Indian J Pathol Microbiol* 2011;54(3):588-590.
2. Doll CD, Weiss RB. Malignant lymphoma of testis. *Am J Med* 1986; 81(3):515-524.
3. D'Abrosca F, Lucev M, Turi V, Gerosa E, Villani L. Primary lymphoma of the spermatic cord. *Minerva Chir* 1990;45(7):531-533.
4. Hautzer NW, Nikolai V. Primary lymphoma of the spermatic cord. *Br J Urol* 1986;58(5):565-566.
5. Lagrange JL, Ramaioli A, Theodore CH et al. Non-Hodgkin's lymphoma of the testis: a retrospective study of 84 patients treated in the French anticancer centres. *Ann Oncol* 2001;12(9):1313-1319.
6. Buskirk SJ, Evans RG, Banks PM, O'Connell MJ, Earle JD. Primary lymphoma of the tesis. *Int J Radiat Oncol Biol Phys* 1982;8(10): 1699-1703.
7. Coiffier, B. Rituximab therapy in malignant lymphoma. *Oncogene* 2007;26(25):3603-3613.
8. Zucca E, Conconi A, Mughal TI et al. Patterns of outcome and prognostic facors in primary large cell lymphoma of the testis in a survey by the International Extranodal Lymphoma Study Group. *J Clin Oncol* 2003;21(1):20-27.
9. Ackland SP, Schilsky RL. High-dose methotrexate: a critical reappraisal. *J Clin Oncol* 1987;5(12):2017-2031.
10. Møller MB. Non Hodgkin's lymphoma of the spermatic cord. *Acta Haematol* 1994;91(2):70-72.