

CASE REPORT

Primary testicular lymphoma in a young male- A rare entity

SANDEEP GUPTA¹, KISHORE KN², VIJAYENDRA KANWAR¹, KHUMUKCHAM SOMARENDRA¹, AKOIJAM KAKU SINGH¹ and SINAM RAJENDRA SINGH¹

¹Department of Urology, ²Department of Surgery, RIMS, Imphal, India.

ABSTRACT

Testicular neoplasms are a diverse group of tumors mostly comprised of germ cell tumors. Lymphoma of the testis are rarely seen, more so in the younger age group. Most of the lymphoma involving the testes is secondary to systemic lymphoma or leukemia. Primary testicular lymphoma, in which there is no systemic evidence of lymphoma or leukemia, is rare. If encountered, it is seen mostly in elderly males. Here we present a case where a middle-aged male was diagnosed with primary testicular lymphoma (diffuse large B cell) without extra-testicular extension..

Key words: Primary testicular lymphoma, diffuse large B cell lymphoma, young.

INTRODUCTION

Primary testicular or para-testicular lymphomas arise in the testicles, epididymis or spermatic cord and are not associated with lymphoma or leukemia elsewhere in the body. Involvement of these anatomic structures by systemic lymphomas/leukemia defines secondary testicular or para-testicular lymphomas.¹

Primary testicular Non-Hodgkin lymphoma is a rare tumor which represents only 1-9% of testicular tumors and 1% of all cases of lymphoma.² Most commonly, lymphoma involves the testis through dissemination from extra testicular sites (secondary testicular lymphoma). But involvement of testis without systemic lymphoma (primary testicular lymphoma) is a rare entity. Even though a rare disease, it is the most common type of testicular neoplasm in elderly males.³ It usually presents as a progressive, painless testicular swelling. These tumors are initially treated with radical inguinal orchidectomy and subsequently radiotherapy and chemotherapy.⁴ Even in this modern era of advanced surgical techniques and radio chemotherapy, the prognosis of primary testicular lymphoma remains poor.⁵

Although few cases have been reported in literature, all of them are in elderly male. Here we report a case of testicular lymphoma we encountered in a middle-aged male at our urology department.

CASE REPORT

A 46-year-old male presented to the urology clinic with scrotal swelling since 4 months, which was gradually increasing in size. There was no history of trauma or pain in the swelling. There were no associated symptoms like burning micturation, fever etc. He also gave a history of decreased sensation over the right hemiscrotum. On clinical examination, there was an approximately 8X6 cm mass in the right side of the scrotum, which was non-tender, having a smooth surface and firm in consistency. Testis couldn't be felt separately where as epididymis was palpated at the superior border of the mass. Ultrasound of inguino-scrotal region showed a hypoechoic lesion in right testis with normal para-testicular tissue. The opposite testis was normal on ultrasound.

His routine investigations like complete hemogram, liver and kidney function test, ECG and specific investigations like chest X-ray and tumor markers like beta-HCG, alpha-fetoprotein, and serum LDH were within the normal range. A Computed Tomography of abdomen and pelvis including scrotum revealed an enlarged right testis with a hypodense lesion measuring 5X5 centimeter in greatest dimension having fluid collection in the scrotal sac (Fig. 1) with normal appearing para-testicular tissue.



Figure 1, CT-scan showing testicular swelling with fluid collection.

Correspondence:

Sandeep Gupta

Department of Urology, RIMS, Imphal, Manipur 795004, India.

Email: drsandeepgupta2009@yahoo.in

The opposite testis was normal. There was no evidence of intra abdominal lymphadenopathy. A diagnosis of right sided malignant testicular tumor was made and the patient underwent high inguinal orchiectomy.

The surgery was uneventful with the excised tumor size measuring 5x5x6 centimeters with a smooth surface (Fig. 2).



Figure 2, Post-operative specimen.

The cut section showed grey white nodular tissue with focal areas of necrosis. Microscopy showed testicular tissue diffusely infiltrated by monomorphic population of lymphoid cells, which were large cells with scanty cytoplasm and pleomorphic vesicular nuclei with distinct nucleoli (Fig. 3).

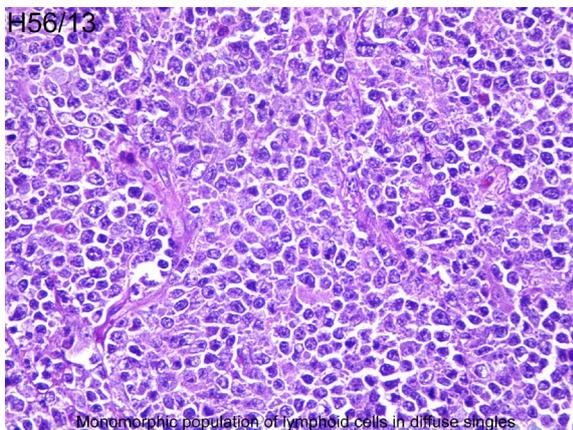


Figure 3, HPE of excised specimen

Many foci of entrapped seminiferous tubules with thickened hyalinised wall were also seen. Tunica vaginalis, epididymis, spermatic cord and soft tissue dissected along with the testis were free from atypical cells. Immuno histochemistry showed CD-20 positivity (Fig. 4-a) and negativity for CD-3 (Fig. 4B).

The tumour was diagnosed as diffuse large B cell lymphoma of the testis and the patient was referred to medical oncology department where he got 6 cycles chemotherapy of R-CHOP (Rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone) regimen and prophylactic involved field radiotherapy of 30Gy over 15 fractions for left testis and 30Gy

over 10 fractions whole brain radiotherapy. Patient is now on regular follow up every 3 months with no signs of recurrence. A repeat CT scan of abdomen and pelvis after 1 year of completing chemo-radiotherapy showed no signs of recurrence.

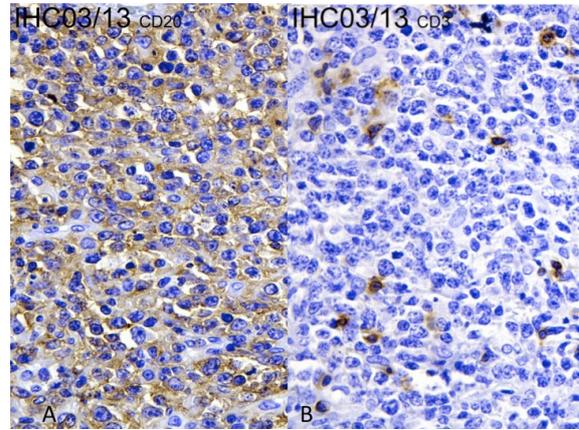


Figure 4a and 4b, Immunohistochemistry with CD-20 & CD-3.

DISCUSSION

Testicular tumor is one of the most common cancer affecting adult male, seminoma being most common histological type.⁶ Among testicular tumor, lymphoma constitutes only 1-9%. Majority of lymphoma arises in lymph nodes and testicular involvement is mostly secondary to systemic disease. But in some cases as in this patient, it occurs primarily in extra nodal sites including the brain, gastrointestinal tract, lungs, skin, breast and testis.³

Primary testicular lymphoma is a condition where lymphoma is found in the testis with no systemic disease or lymphoma in any other organ system. Malassez et al reported the first case of testicular lymphoma in the year 1877. Most of these are B cell origin, 85% constituting diffuse large B cell lymphoma (DLBC), which are intermediate or high-grade.⁶ These affect adult males with mean age being 60 years. There is no side predominance (left = right) and bilateral involvement is seen rarely (6%) except in lymphoblastic lymphoma in which simultaneous bilateral involvement is characteristic.⁵

The risk factors for testicular lymphoma are immunosuppression, chronic orchitis, trauma and filariasis.¹¹ Most of the patients present themselves in stage I (56%),⁷ the commonest mode of presentation being painless testicular swelling. Constitutional symptoms like fever, weight loss and night sweat are seen in 25 to 40% of the patients. Patients presenting without B symptoms are not rare, as our patient had no B symptoms.

These tumors may infiltrate locally and cases involving epididymis, spermatic cord and scrotal skin have been reported⁹ but in our case no such local infiltration by tumor cells was noted. Metastases to distant sites can also be seen, most commonly in central nervous system, Waldeyer's ring, lung, prostate and

renal system. Ultrasound of inguino-scrotal region and computed tomography of abdomen will aid in the diagnosis and staging of the tumour.

Bad prognosis is associated with higher patient age, poor performance status, advanced stage or more bulky disease, higher LDH, bone marrow involvement. The primary (stage IE) lymphomas of the testis and spermatic cord have the worst prognosis among all extranodal lymphomas, with 5 year overall survival rates of 70-79%.

High inguinal orchidectomy still remains the initial procedure of choice for diagnosis and management. Before the advent of radiotherapy, patients who underwent high inguinal orchidectomy alone had poor 5-year survival rate (10-12%). After 1980 when postoperative radiotherapy was introduced, the 5-year survival increased to 64% in stage 1 disease.⁷ Addition of rituximab to previous CHOP (R-CHOP) regime has shown promise in 5-year survival rate.⁴ Prophylactic radiotherapy for contralateral testis and CNS is advised.

CONCLUSION

Primary testicular lymphoma is a rare but aggressive tumour, which presents as a painless unilateral testicular swelling. These tumors are seen most commonly in elderly males but it should be kept in mind when encountered with a young patient having painless testicular swelling. High inguinal orchidectomy, chemotherapy with prophylactic radiotherapy to contralateral testis and CNS is the treatment. Still primary testicular lymphoma has the worst prognosis among lymphoma.

REFERENCES

- Miedler JD, MacLennan GT. Primary testicular lymphoma. *J Urol*. 2007; 178: 2645.
- Moller MB, d'Amore F, Christensen BE. Testicular lymphoma: A population-based study of incidence, clinic-pathological co relations and prognosis. The Danish Lymphoma Study Group, LYFO. *Eur J Cancer*. 1994; 30A: 1760-64.
- ShahabN, Doll DC: Testicular lymphoma. *Semin Oncol*. 1999; 26:259-69.
- Umberto Vitolo, Annalisa Chiappella, Andrés J.M. Ferreri, et al: First-Line Treatment for Primary Testicular Diffuse Large B-Cell Lymphoma With Rituximab-CHOP, CNS Prophylaxis, and Contralateral Testis Irradiation: Final Results of an International Phase II Trial. *J Clin Oncol*. 1995; 13:1361-67
- Darby S, Hancock BW. Localised Non-Hodgkin lymphoma of the testis: the Sheffield Lymphoma Group experience. *Int J Oncol*. 2005; 26:1093-95.
- Hasselblom S, Ridell B, Wedel H, Norrby K, Sender Baum M, Ekman T. Testicular lymphoma, a retrospective, population based, clinical and immuno-histo-chemical study. *Acta Oncol*. 2004; 43:758-65.
- Zietman AL, Coen JJ, Ferry JA, Scully RE, Kaufman DS, McGovern FG. The management and outcome of stage IAE non-Hodgkin's lymphoma of the testis. *J Urol*. 1996; 15:943-46.
- Seymour JF, Solomon B, Wolf MM, Januszczyk EH, Wirth A, Prince HM. Primary large-cell non-Hodgkin's lymphoma of the testis: A retrospective analysis of patterns of failure and prognostic factors. *Clin Lymphoma*. 2001; 2:109-15
- Ferry JA, Harris NL, Young RH, Coen J, Zietman A, Scully RE. Malignant lymphoma of the testis, epididymis, and spermatic cord: A clinicopathologic study of 69 cases with immunophenotypic analysis. *Am J Surg Pathol*. 1994; 18:376-90
- Fonseca R, Habermann TM, Colgan JP, et al. Testicular lymphoma is associated with a high incidence of extranodal recurrence. *Cancer*. 2000; 88:154-61
- Sokovich RS, Bormes TP, McKiel CF. Acquired immunodeficiency syndrome presenting as testicular lymphoma. *J Urol*. 1992; 147:1110-11.
- Zucca E, Conconi A, Mughal TI, et al. Patterns of outcome and prognostic factors in primary large-cell lymphoma of the testis in a survey by the International Extranodal Lymphoma Study Group. *J Clin Oncol*. 2003; 21:20-27