

Case Report

Testicular non-Hodgkin's lymphoma – a rare tumor

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ABSTRACT

Primary lymphoma of the testis is an exceedingly rare disease. We present a case of a 65 years old gentleman who presented with a brief history of testicular pain. Imaging studies and serum tumour markers indicated a testicular lesion of suspicious aetiology. High inguinal orchidectomy was performed. Histopathology and immunohistochemistry revealed diffuse large B-cell lymphoma. Positron emission tomography (PET) scan revealed a metabolically active retroperitoneal lymph node in aortocaval location. Subsequently he underwent chemotherapy with Rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone (R-CHOP) regimen plus intravenous Methotrexate, following which PET scan showed disappearance of the previously detected metabolically active lesion.

Keywords: Non-Hodgkin's lymphoma, Testis

INTRODUCTION

Primary testicular lymphoma (PTL) is a rare disease with an annual incidence of 0.26 cases per 100,000.¹ It is the most common testicular malignancy in elderly males above the age of 60 years, with the most common histological subtype being diffuse large B-cell lymphoma (DLBCL).^{2,3} Clinical suspicion of a testicular neoplasm can be supported by imaging studies and estimation of serum tumor marker levels. However final diagnosis can be made only after histological studies. Treatment protocol includes a high inguinal orchidectomy followed by chemotherapy.

Here we present a case of a PTL that was managed at our centre.

CASE REPORT

The patient, a 65 years old gentleman, presented with complaints of right testicular pain for 3 days. Examination revealed an enlarged, non-tender right testis with loss of testicular sensation. There was no palpable inguinal lymphadenopathy and no palpable abdominal lump. There

was no significant finding on examination of spine, cervical lymph nodes and respiratory system.

Ultrasonogram of the scrotum was performed, which gave differential diagnoses of orchitis or testicular neoplasm. Serum markers for testicular cancer were assayed, which were reported as alpha fetoprotein (AFP) 5.11 ng/ml (reference range 0-9.0), β -human chorionic gonadotropin (HCG) 0.40 mIU/ml (reference range <0.5-2.67 for males) and lactate dehydrogenase (LDH) 302 U/L (reference range <248). A contrast enhanced computed tomography (CT) scan of the abdomen was done, which showed a "bulky right testis with heterogeneous enhancement and mild hydrocele with varicocele." The patient was advised to undergo a right sided high inguinal orchidectomy in view of the testicular mass suspicious of neoplasm and the elevated LDH levels. The surgery was performed under spinal anesthesia and the testis with spermatic cord was sent for histopathological examination. His post-operative period was uneventful.

The histopathology examination was reported as non-Hodgkin's lymphoma (Figure 1-3). He was then referred to an oncology center, where the immunohistochemistry

tests showed that the tumor cells were immunopositive for CD 20 and MUM 1, and negative for CD 3, CD 10, BCL 6 and cyclin D1. The final impression was that of diffuse large B-cell lymphoma NOS, post germinal centre type. Subsequently he underwent a whole-body positron emission tomography (PET-CT) scan and a contrast enhanced computed tomography (CT) scan of the brain. A metabolically active lymph node (?mitotic) was detected in the retroperitoneum (aortocaval – L3 level) measuring 1.4 cm. Bone marrow biopsy was done which did not show any tumour cells.

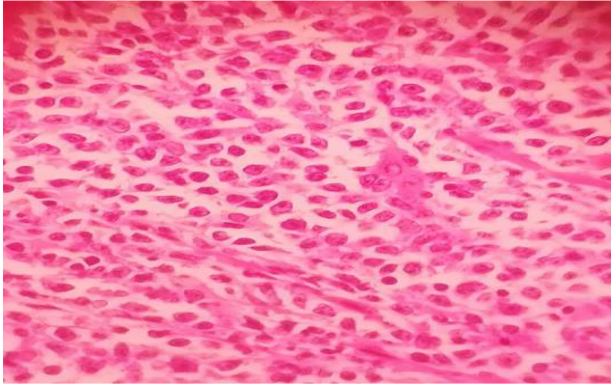


Figure 1: Tumor cells at 40X magnification.

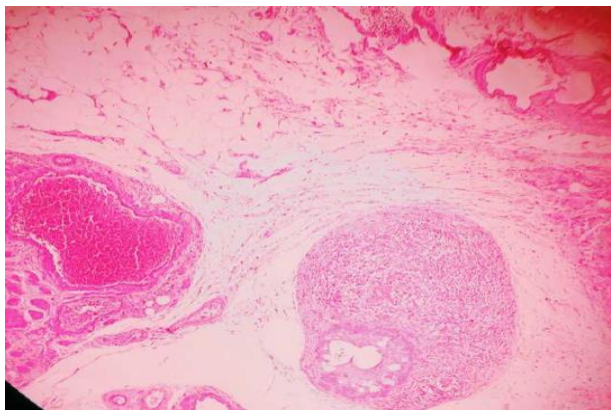


Figure 2: Spermatic cord with tumour infiltration.

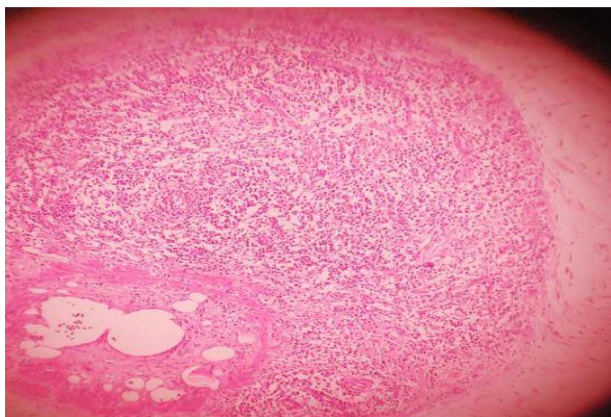


Figure 3: Spermatic cord with tumour infiltration (magnified).

He received 6 cycles of chemotherapy with rituximab, adriamycin, cyclophosphamide, vincristine and prednisolone (R-CHOP regimen), along with intravenous methotrexate.

Upon completion of chemotherapy, whole-body PET-CT scan was repeated, which showed disappearance of the previously detected metabolically active node and no new lesions were seen.

Currently the patient is doing well and is on regular follow-up. Post operatively the lactate dehydrogenase (LDH) levels had normalized and he currently awaiting PET scan after completion of 1 year after chemotherapy.

DISCUSSION

Testicular lymphoma is a rare disease with an annual incidence of 0.26 cases per 100,000 and the most common testicular malignancy in elderly males above the age of 60 years.^{1,2} This accounts for about 1% to 2% of all non-Hodgkin lymphomas (NHLs) and approximately 5% of all testicular neoplasms.² The most common histological subtype of PTL is diffuse large B-cell lymphoma (DLBCL).³ Testicular lymphoma carries a poor prognosis compared to other NHL and extranodal lymphomas and may require a more prolonged course of chemotherapy compared to other extranodal lymphomas.^{4,5}

The first step of management of a suspicious testicular neoplasm is an orchidectomy. There is no definite consensus regarding further management, due to the rarity of the disease and available literature for the same.² In an article by Connors, it was recommended that patients with early-stage disease IAE/IIAE should receive brief chemotherapy with involved field radiation including the contralateral testis.⁶ However our patient did not receive radiotherapy as per the consensus of the multidisciplinary team. Patients with primary testicular involvement have a particularly high risk of central nervous system (CNS) involvement (>15%) and treatment recommendations for these patients differ from those for other forms of extranodal DLBCL.⁷ A study by Vitolo and colleagues reported a 6% CNS relapse rate after 5 years in patients treated with a combination of R-CHOP plus four doses of intrathecal methotrexate and contralateral testis irradiation.⁸ Our patient had received intravenous methotrexate for CNS prophylaxis. Because of the poor prognosis, an aggressive treatment approach is warranted. However, testicular lymphoma is predominantly a disease of older men who often have limited ability to tolerate aggressive treatment.

The ideal management protocol for patients with testicular lymphoma is yet to be formulated.

CONCLUSION

PTL is an uncommon disease, usually affecting the elderly. Treatment includes surgical removal of the testis (which

gives histological diagnosis), followed by chemotherapy and radiation. The regimens have to be individualised depending on the disease stage patient's general condition.

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