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RESEARCH ARTICLE

Primary Testicular involvement of Nonhodgkins Lymphoma.

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Abstract

Primary testicular NHL is very rare disease accounts only 1-2% of NHL. It is important to identify primary testicular lymphoma correctly and distinguish it from other entities because of difference in therapy, management and prognosis.

INTRODUCTION

Primary non-Hodgkin's lymphoma of the testis (PTL) accounts for about 5% of testicular neoplasm and 1–2% of all non-Hodgkin's lymphomas. It is the most common testicular malignancy in elder men¹ Testicular lymphoma has a higher incidence of bilateral involvement. Intermediate-grade diffuse large B-cell lymphoma is the most common histological pattern among primary testicular lymphoma, Although excellent results with a doxorubicin-containing chemotherapy regimen have been achieved in early-stage disease, patients with advanced disease have a grave prognosis.²

Case Report-

A 59 yrs old male patient presented with swelling in right scrotal region since 6 month which was gradual increase in the size and also has dragging pain in Right scrotal region since 1 month. There was no h/o trauma, wt.loss, T.B,D.M.

On clinical examination there was swelling of size 15 x 10 cm present in the rt scrotal region , non tender, no cough impulse seen & one can get above the swelling. Fluctuation & Transillumination test are positive. Testis not palpable.

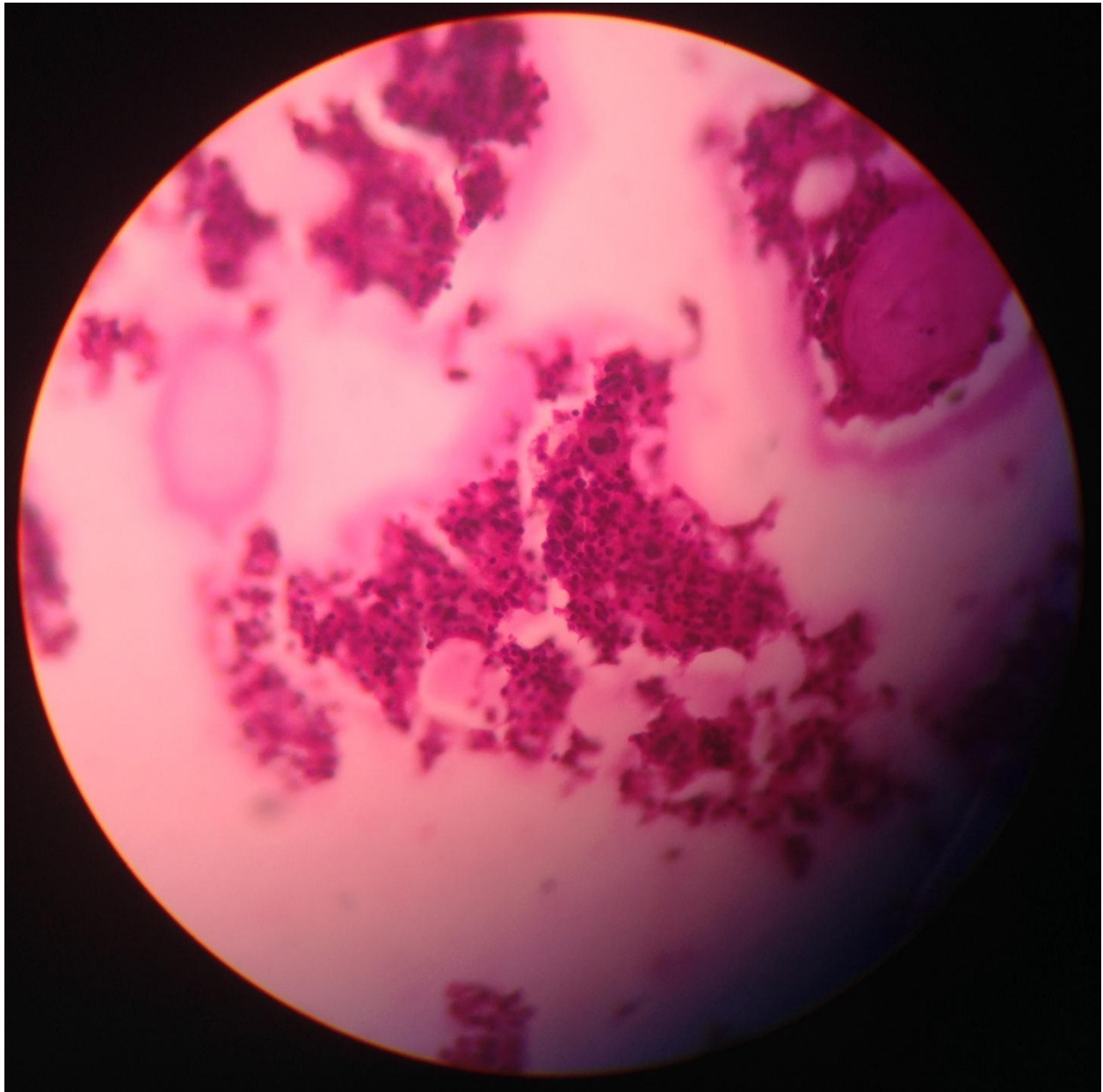
The clinical diagnosis was **Right Vaginal Hydrocele**.

The patient was investigated,

USG scrotum showed- both testis are slightly enlarged & normal in echotexture. Bilateral simple hydrocele more on right side about 200ml than left side of about 15 ml. so patient was investigated & subjected to right Eversion of sac.

Operative finding- on right side there was 200 ml yellowish fluid was present , testis was enlarged & more vascular. Fluid was drained. Decision was made for testicular biopsy & eversion of sac.

Testicular biopsy (Histopathological finding)- Non Hodgkin Lymphoma of Diffuse large B cell type. The tumour cell express CD 20 , Bcl2 & BCL6C (weakly & focally). They are Immunonegative for CD3 & CD5. The mib – 1 proliferation index is approximately 80 %.(fig 1).



Bone marrow aspiration was within normal limit.

CT scan Neck,chest,abdomen & pelvis were within normal limit.

Impression – The studies are suggestive of primary testicular involvement of Non Hodgkin Lymphoma. Patient was started on intrathecal methotrexate &R- CHOP regime.

Discussion –

DLBCL was the commonest primary testicular lymphoma. Testicular lymphoma is a rare and deadly form of extra nodal lymphoma. It is a rare disease and accounts for 1% of all non-Hodgkin's lymphoma (NHL) cases, 2% of all extra nodal lymphomas and 5% of all testicular neoplasms.¹⁰. TNHL usually occurs in older men and is the most common testicular malignancy in men between sixty and eighty years of age. High incidence of bilateral testicular

involvement, which is a unique feature of TNHL, was first reported by Abeshouse et al. in 1955.² TNHL is now considered to be the most common bilateral tumor of the testes,^{2,4} with reported incidence of bilateral metachronous testicular involvement of 35% and bilateral synchronous testicular involvement of 3%.⁴ Other authors have reported the incidence of bilateral involvement of about 10-40%.^{11,7,9.}

It is important to identify primary testicular lymphoma correctly and distinguish it from other entities because of difference in therapy and prognosis. There are neither any well-documented etiological or predisposing factors nor any significant associations existing between histories of trauma, chronic orchitis or cryptorchidism and subsequent development of TNHL.^{2,3,4,5} TNHL has a predilection for dissemination to non-contiguous extranodal sites such as the CNS, Waldeyer's ring, skin and lungs.^{2,3,6,7} Martenson et al. reported a 10% incidence of CNS involvement in the initial failure and 20% in subsequent relapses.⁸ Thus, lumbar puncture is warranted as one of the initial procedures at the time of diagnosis.^{3,15} According to the Working Formulation of the United States National Cancer Institute, approximately 68% of TNHL cases are classified as intermediate grade, diffuse large B-cell subtype, followed by high-grade, diffuse small non-cleaved subtype in about 30% of the patients.^{2,7} There is no prognostic advantage for any pathological subtype.⁷ Immunohistochemistry (IHC) studies confirm the majority of TNHL cases to be of B-cell origin, with lesser occurrence of T-cell lymphoma.^{2,12} Histopathological differentiation of TNHL from seminoma is usually a challenge.⁴ Other conditions that might resemble TNHL are embryonal cell carcinoma, granulomatous orchitis, pseudolymphoma, plasmacytoma and rhabdomyosarcoma.^{2,4,13} Serum lactate dehydrogenase (LDH) levels have been correlated with tumor aggressiveness,¹² whereas other tumor markers such as serum beta human chorionic gonadotropin (HCG) and serum alpha-fetoprotein (AFP) are rarely elevated in TNHL cases.¹² Al-Abbadi et al. studied 18 patients and classified their disease as primary testicular lymphoma with germinal center B-cell-like and non-germinal center B-cell-like by means of the IHC expression of CD10, Bcl-6 and MUM1. They found that 89% of the primary testicular lymphoma of the diffuse large B-cell type belonged to the non-germinal center B-cell-like subgroup and all exhibited high proliferative activity. The germinal center B-cell-like type of primary testicular lymphoma was uncommon and was seen mostly in HIV-positive patients.¹⁴ Treatment is complex and dependant on the initial characteristic of patient.

It is important to identify primary testicular lymphoma correctly and distinguish it from other entities because of difference in therapy, management and prognosis. The high incidence of spreading, especially to the CNS, leads to advocacy of the use of CNS prophylaxis with intrathecal chemotherapy. Prospective multicenter trials incorporating a large number of patients would lead to better treatment options for this subtype. For stages IE and IIE, there is universal agreement on orchidectomy as the initial treatment. If treated with orchidectomy alone, the majority of these patients relapse within the first two years at various extranodal sites, and hence the use of adjuvant systemic chemotherapy, radiotherapy and also prophylactic intrathecal chemotherapy has been emphasized.^{6,15} For stage IIIIE and IVE disease, the treatment of choice is systemic chemotherapy, with irradiation reserved for symptomatic and bulky localized deposits.⁷ Although the majority of patients achieve complete remission, most of them relapse, with a median survival of three to five months.⁶ In view of the high risk of CNS relapse, even in patients who achieve complete remission with primary therapy, CNS prophylaxis should be considered in all advanced-stage patients.¹² Similarly, prophylactic irradiation for the contralateral testis is recommended, since the relapse rate in the contralateral testis is up to 50%.¹⁰ Nevertheless, contrary to this approach, some authors argue against prophylactic radiotherapy for the contralateral testis.¹⁶

Conclusion –

If the patient come with Hydrocele and having testicular enlargement then testicular biopsy is most important to rule out the rarest disease. It is important to identify testicular diseases correctly and distinguish it from other entities because of difference in therapy, management and prognosis.

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