

Primary testicular diffuse large B-cell lymphoma in young patient: an uncommon presentation

Abstract

Primary testicular lymphoma (PTL) is a rare form of extranodal lymphoma, this neoplasm is the most common malignant tumor of the testis in the elderly age, and the majority of cases are histologically diffuse large B-cell lymphoma (DLBCL), accounting for 80% to 98% of non-hodgkin lymphoma. Patients with primary testicular DLBCL show a continuously high risk of recurrence with no plateau in the survival curves and a tendency to involve other extranodal sites, especially the central nervous system and the contralateral testis.

We report a rare and uncommon presentation of a primary testicular diffuse B cell lymphoma in young patient.

Keywords: primary testicular lymphoma, diffuse large B-cell lymphoma, testicular mass

Volume 10 Issue 4 - 2022

Alami Drideb, Bouchnafat, Saddi

Internal medicine and Oncohematology department, HASSAN II University hospital, Sidi Mohammed Ben Abdellah University, Morocco

Correspondence: Alami Drideb, Internal medicine and Oncohematology department, HASSAN II University hospital, Sidi Mohammed Ben Abdellah University, Morocco, Tel 00212623496994, Email noufissa.alami@gmail.com

Received: November 14, 2022 | **Published:** December 13, 2022

Introduction

Primary testicular lymphoma (PTL) is an uncommon and aggressive form of extranodal testicular diffuse large B-cell lymphoma (TDLBCL) in young population, accounting for 1 to 2% of Non-Hodgkin Lymphoma cases.¹ We report a very rare case of a primary testicular diffuse B cell lymphoma in young patient.

Case report

A 30 year's old men, with a six months history of an isolated left testicular mass of 7 cm with hard consistency, no inflammatory signs and no lymphadenopathy. The scrotal ultrasound and CT scan found a large heterogeneous left testicular mass infiltrating the skin and the penile root, it measures 60 x38 mm (Figure 1). Left orchiectomy was performed and histopathology showed non germinal center diffuse large B cell lymphoma: CD20+, CD3-, Ki67 at 80%, CD10-, CD30-, cytokeratin-, PLAP-, Alpha-fetoprotein-, and CD117-. Full body computed tomography indicated some lumbo-aortic and latero-aortic adenopathy, measuring for the largest 17mm of the small axis. No other lymph nodes or spleen abnormality. The bone marrow biopsy was normal. Serum lactate dehydrogenase (LDH) level was at 471 IU/L, beta2-microglobulin at 5.4 mg/dL and uric acid at 9,4 mg/dl, B and C hepatitis serology and HIV screening were normal. The patient was classified stage II E with prognostic 2 (according to the International Prognostic Index (IPI), and low to intermediate risk group. Treatment protocol was RCHOP (Rituximab, Cyclophosphamide, Doxorubicin Vincristine Prednisolone), in addition to four prophylactic intrathecal administrations of Methotrexate.



Figure 1 Injected time axial image showing testicular mass.

He achieved a partial response after four cycles of the RCHOP protocole, and the treatment was continued. After the sixth cycle, he developed a 7 cm scrotal skin ulceration with infiltrated base and fibrinous surface. The biopsy confirmed a tumor proliferation of large cell B-cell lymphoma. A 18 FDG PET-Scan showed intense hypermetabolic left flank mass (SUV 10, 6) and left scrotal fixation mass (SUV at 9.1) of 10 cm diameter (Figure 2).

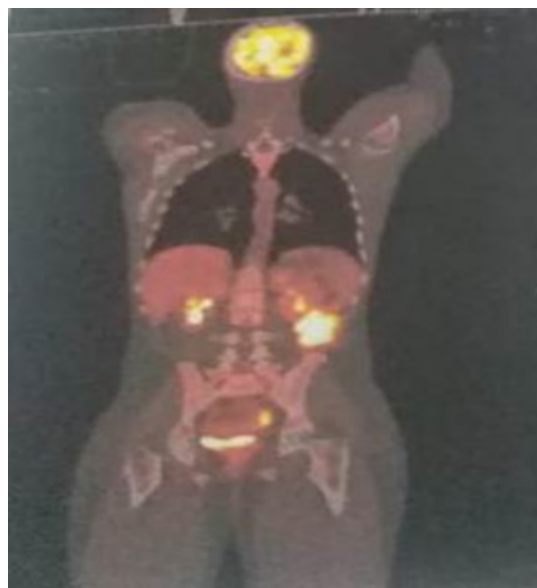


Figure 2 FDG PET intense hypermetabolic left flank mass and left scrotal fixation mass.

Discussion

TDLBCL is a lymphoid malignancy of the B-cells and it is the most common type of non-Hodgkins lymphomas affecting the testis. Age at diagnosis vary between 60 and 70 years.² Most cases are classified as a non-germinal center cell-of-origin subtype, which may partially account for the aggressive nature of the disease.³ The tumor microenvironment (TME) of T-DLBCL has been demonstrated to harbor characteristic features that highlight the role of immune escape.⁴ The patients with T-cell inflamed TME had a significantly increased risk of progression and death independently of IPI. Furthermore,

reduced membranous staining of HLA I and II correlated with low T-cell infiltration.³ HIV infection is a recognized risk factor for aggressive and primary extranodal lymphomas, well-described factor of T-DLBCL,⁵ but the etiology remains largely unknown. T-DLBCL typically presents as a painless mass, and has a high tendency to disseminate to the contralateral testis and central nervous system (CNS), but also to other extranodal sites, such as lung, pleura, skin, soft tissues and Waldeyer's ring.⁶ Approximately 20% Have locally advanced stage II disease.⁵

The outcome of T-DLBCL patients can be estimated according to the IPI, or the modified age-adjusted IPI, revised IPI, or the National Comprehensive Cancer Network IPI.⁷ In addition to the complete remission, the Purpose of the treatment avoids relapses of the contralateral testis and the CNS. The internationally recognized standard of care consists of orchiectomy followed by immunochemotherapy with six cycles of R-CHOP or R-CHOP-like Regimen given every 21 days.^{8,9} The addition of CNS prophylaxis with IV administered CNS-penetrating chemotherapy such as high dose methotrexate (HD-Mtx) or high Dose cytarabine (HD-Ara-C) and/or IT chemotherapy as well as irradiation or excision of The contralateral testis are highly recommended.^{8,9}

Conclusion

This paper reports the only patient diagnosed with primary testicular diffuse B cell lymphoma in the onco Hematology and internal medicine department in the last 10 years. T-DLBCL remains a rare extra nodal lymphoma but with an aggressive clinical course. Clinical trials testing novel drugs are an interesting area of future research, especially for patients who failed this strategy or who relapse.

Acknowledgements

None

Conflicts of interest

The author declares no conflicts of interest.

References

1. Møller MB, Amore F, Christensen BE. Testicular lymphoma: a population-based study of incidence, clinicopathological correlations and prognosis. The Danish Lymphoma Study Group, LYFO. *Eur J Cancer*. 1994; 30A(12):1760–1764
2. Mannisto, Vahamurto, Pollari, et al. Intravenous but not intrathecal central nervous system-directed chemotherapy improves survival in patients with testicular diffuse large B-cell lymphoma. *Eur J Cancer*. 2019;115:27–36.
3. Leivonen, Pollari, Bruck O, et al. T-cell inflamed tumor microenvironment predicts favorable prognosis in primary testicular lymphoma. *Haematologica*. 2019;104(2):338–346.
4. Pollari, Bruck O, Pellinen, et al. PD-L1+ tumor-associated macrophages and PD-1+ tumor-infiltrating lymphocytes predict survival in primary testicular lymphoma. *Haematologica*. 2018;103(11):1908–1914.
5. Pollari M, Leppä S. Testicular Diffuse Large B-Cell Lymphoma—Clinical, Molecular, and Immunological Features. *Cancers*. 2021;13(16):4049.
6. Mazloom A, Fowler N, Medeiros LJ, et al. Outcome of patients with diffuse large B-cell lymphoma of the testis by era of treatment: the M. D. Anderson Cancer Center experience. *Leuk Lymphoma*. 2010;51(7):1217–1224.
7. Zhou Z, Sehn L.H, Rademaker, et al. An enhanced International Prognostic Index (NCCN-IPI) for patients with diffuse large B-cell lymphoma treated in the rituximab era. *Blood*. 2014;123(6):837–842.
8. NCCN. Clinical Practice Guidelines for B-Cell Lymphomas.
9. Vitolo U, Seymour J F, Martelli M, et al. Extranodal diffuse large B-cell lymphoma (DLBCL) and primary mediastinal B-cell lymphoma: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol*. 2016;27suppl(5):v91–v102.