

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

N.ALAMI DRIDEB, S.BOUCHNAFATI, N.SADDIQ, K.MELIANI,
S.SAOUDI, M.OUAZZANI, Z.KHAMMAR, R. BERRADY.

Internal medicine and oncohematology department, HASSAN II university hospital, Sidi Mohammed Ben Abdellah University, Fez, Morocco

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

Date of Submission: 19-11-2022

Date of Acceptance: 03-12-2022

I. 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 and for 5% of testicular malignancies [1]. We report a very rare case of a primary testicular diffuse B cell lymphoma in young patient.

II. 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 orchietomy 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, DoxorubicinVincristine Prednisolone), in addition to four prophylactic intrathecal administrations of Methotrexate.

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).

III. 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) has an important role in lymphoma development. 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 International Prognostic Index 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].

IV. 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.

References

- [1]. Møller MB, d'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, S.; Vahamurto, P.; Pollari, M.; Clausen, M.R.; Jyrkkio, S.; Kellokumpu-Lehtinen, P.L.; Kovanen, P.; Karjalainen-Lindsberg, M.L.; d'Amore, F.; Leppä, S. 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, S.K.; Pollari, M.; Bruck, O.; Pellinen, T.; Autio, M.; Karjalainen-Lindsberg, M.L.; Mannisto, S.; Kellokumpu-Lehtinen, P.L.; Kallioniemi, O.; Mustjoki, S.; et al. T-cell inflamed tumor microenvironment predicts favorable prognosis in primary testicular lymphoma. *Haematologica* 2019, 104, 338–346.
- [4]. Pollari, M.; Bruck, O.; Pellinen, T.; Vahamurto, P.; Karjalainen-Lindsberg, M.L.; Mannisto, S.; Kallioniemi, O.; Kellokumpu-Lehtinen, P.L.; Mustjoki, S.; Leivonen, S.K.; et al. PD-L1+ tumor-associated macrophages and PD-1+ tumor-infiltrating lymphocytes predict survival in primary testicular lymphoma. *Haematologica* 2018, 103, 1908–1914.
- [5]. Pollari, M.; Leivonen, S.-K.; Leppä, S. Testicular Diffuse Large B-Cell Lymphoma—Clinical, Molecular, and Immunological Features. *Cancers* 2021, 13, 4049
- [6]. Mazloom A, Fowler N, Medeiros LJ, Iyengar P, Horace P, Dabaja BS. 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): 1217-24.
- [7]. Zhou, Z.; Sehn, L.H.; Rademaker, A.W.; Gordon, L.I.; Lacasce, A.S.; Crosby-Thompson, A.; Vanderplas, A.; Zelenetz, A.D.; Abel, G.A.; Rodriguez, M.A.; 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, 837–842.
- [8]. NCCN. Clinical Practice Guidelines for B-Cell Lymphomas. Available online: <https://www.nccn.org/guidelines/guidelinesdetail?category=1&id=1480> (accessed on 22 June 2021)
- [9]. Vitolo, U.; Seymour, J.F.; Martelli, M.; Illerhaus, G.; Illidge, T.; Zucca, E.; Campo, E.; Ladetto, M.; Committee, E.G. 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, 27, v91–v102

N.ALAMI DRIDEB, et. al. "Primary testicular diffuse large B-cell lymphoma in young patient: an uncommon presentation." *IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, 21(11), 2022, pp. 31-32.