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Case Report

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Diagnosis and treatment strategies for primary testicular lymphoma in patients with monorchidism: A case report

Kharbach Youssef[1,2], Tristan Gateau[1], Moulay Hassan Farih[2], Francois Junès[1]

[1]Department of Urology, Robert Boulin Hospital, 33500 Libourne, France

[2]Department of Urology, Hassan II University Hospital, Fez, Maroc

[*]**Corresponding author:** Youssef K, Department of Urology, Hassan II University Hospital, Route de SidiHarazem, 30000 Fez, Morocco, Tel: (+212) 6 62 19 89 00; E-mail: youssef.kharbach@usmba.ac.ma

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Abstract

Monorchidism is relatively frequent. Our case highlights the possibility to be conservative for testicular tumors in monorchid patients while meeting the oncologic objectives.

We report the case of a 52-year-old patient with a history of diffuse large B-cellLymphoma of the test is diagnosed after having an orchiectomy for a unilateral testicular tumor. This patient presented with a testicular lump associated with a hydrocele. Surgical biopsy revealed a non-Hodgkin's lymphoma. No other locations of this tumor was found. The patient had only chemotherapy. Orchiectomy was not performed.

Our case presentation explains how testicular biopsy can be practical in amonorchid with the aim of assuring test is conservation.

Keywords: Lymphoma; Testis; Treatment; Monorchid

Background

Diffuse large B-cell Lymphoma of the testisis rare. Its treatment depends on age, patient medical state and patient's past treatments. We report the case of a patient with a history of orchiectomy for a testicular tumor of which his to pathological examination found a diffuse large B-cell lymphoma. He presented with a testicular lump associated with a hydrocele on the remaining testis. We will describe our diagnostic and therapeutic strategy that was considered in order to preserve the testis whilst meeting the oncologic objectives.

Case presentation

We report the case of a 52-year-old French man with history of stage IV diffuse large B-cell lymphoma of the right testis after under going orchiectomy of the right testis 10 years ago. CT scan objectified an enlarge dright lateral aortic lymphnode (4cm in diameter). The patient received four cycles of R-CHOP (rituximab, cysclophosphamide, doxorubicin, vincristine and prednisolone) with an evaluationat the end of treatment that showed a complete remission on CT scan. The patient had supported chemotherapy very well. Subsequently, the patient under went radiotherapy on all the paraaortic and iliaclymph nodes as well as on the scrotum (30 Gy) with an exta 10 Gy focalized on the right lateralaortic lymphnodes.

Regular clinical and biological monitoring was unremark able during a period of 10 years until the patient presented a progressive swelling on the left test is which had begun two months earlier.

Clinical examination found a patient in good medical state, a s lightly painful testicular lump. No other mass was found after a

complete physical exam was done.

An ultrasound scan was performed which showed a hetero geneous testicular hypertrophy associated with hydrocele.

We were the refore confronted with three possibilities:

1. Testicular Germ cell tumor.
2. B-cell lymphoma relapse.
3. « De novo » lymphoma.

Serumtumor markers were all negative.

Given the patient'shistory of orchiectomy and the in ability to perform fine-needle aspiration due to the hydrocele and the risk of tumor cell dissemination coupled with the patient's refusal to undergo orchiectomy on his remaining testis,we opted for a surgical testicular biopsy which revealed the presence of a diffuse large B- cell lymphoma of the testis (DLBCL) expressing BCL2 and BCL6. Biomolecular analysis using interphase FISH studies detected no rearrangement on the locus of the MYC gene. Cytology of the hydrocele fluid was negative (Figure No : 1, 2, 3).

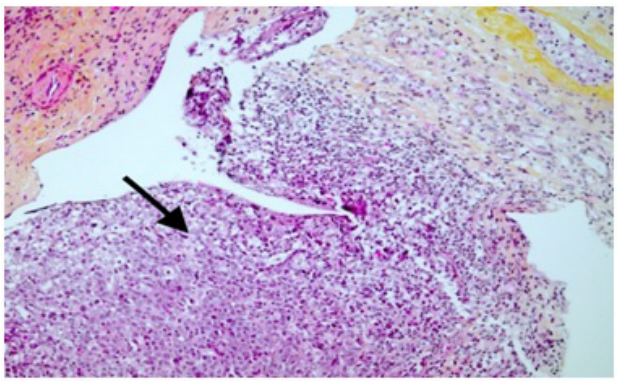

Figure No 1: Histological appearance showing a lymphomatous proliferation containing large polymorphous cells with in the testicular parenchyma (x100) (Arrow: lymphomatous proliferation).

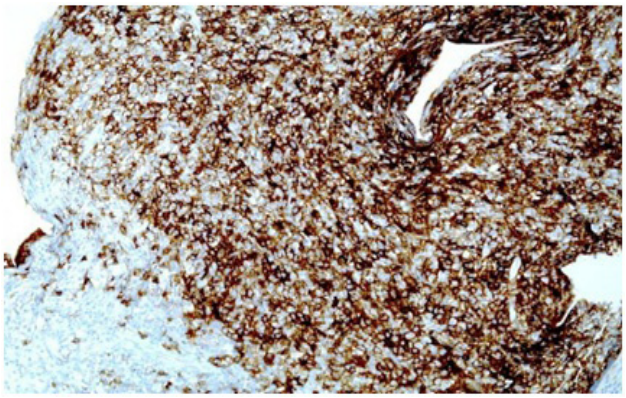

Figure No 2: Histological appearance showing expression of CD20 in immuno histochemistry (x100).

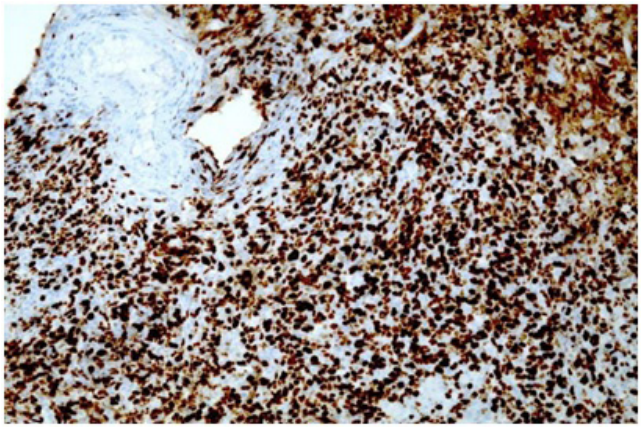

Figure No 3: Histological appearance after using the MIB-1 (x100).

Given these findings we performed tumor staging:

1. Bone marrow biopsy: Showed no invasion.
2. Lumbar puncture: showed an a cellular cerebrospinal fluid.
3. PET scan: Found a hypermetabolic necrosis isolated in the left testicle with no other locations. We how ever retained the diagnosis of a stage IV diffuse large B cell lymphoma of the test is.

The patient received chemotherapy with 4 cycles of R-CHOP + 4 therapeutic lumbar punctures associated with Methotrexate.

Tumor evaluation at the end of treatment found a complete remission on both PET scan and thoraco-abdominal and pelvic CT scan.

Discussion

Primary testicular non-Hodgkin lymphoma (NHL) is exceptionally rare with an annual incidence of 0.26 / 100,000 [1]; 85% of these lymphomas are diagnosed after the age of 60 years[2] and very often after orchiectomy [3].

The peculiarity of our patient already resides in the young age of discovery with significant delay of relapse (10 years) which was limited to the testis.

Risk factors for primary testicular lymphoma are not well known; chronic orchitis, cryptorchidism, trauma or filariasis have been implicated in some cases [4].

Diffuse large B cell lymphomais the most common histological type [4]. The average age of discovery is between 60 and 69 years [5]. And most publications on testicular lymphomas correspond to isolated cases [1].

The DLBCL isoften in the form of a rapidly progressive unilateral testicular hypertrophy [6]. Systemic signs may associate fever, anorexia, night sweats and recent weight loss; they are present

in about 25 to 41% of cases [1].

Tumor invasion of the scrotum and regional retroperitoneal lymphnodes often occurs with this pathology [6]. Local invasion to the epididymis, spermaticcord and scrotal skin may been countered and hydrocele may be found in 40% of cases [6].

Ultrasound shows an intratesticular mass of low echogenicity compared to normal testicular tissue or an enlarged hypoechoictesticle [1].

Stagingis based on Ann Arbor's staging[7]. DLBCL of the testis is known to extend to the opposite testis, Waldeyer's ring, skin, lungs and central nervous system [2]. Serumlevels of tumor markers in testicular tumors: HCG and alpha-FP are rarely high. However, LDH: a marker for assessing tumor size may have a role to monitor the response to therapy [6].

On the histological level, tumor cells often penetrate the tissue spaces diffusely but usually with out destroying the basic architecture [6]. They express the markers usually found in B lymphocytes such as CD19, CD20, CD22 and CD79a but some times one or more of these markers may be lacking [8]. Immuno histochemical study permits to distinguish sub-types of testicular DLBCL based on the presence of CD10, BCL6 and MUM-1 and to differentiate between germinal center B-cell-like DLBCL and not germinal center B- cell-like. This classification has a prognostic value [5-9].

Tumor staging similar to other lymphomas in cludes a thoraco-abdominal and pelvic CT scan, a PET scan and a bone marrow biopsy. The practice of lumbar punctureis recommended for the significant risk of extension to the central nervous system [9].

Because of the rarity of this entity, there is no consensus for treatment [10]. Reported Therapeutics trategies for primary testicular lymphoma associates orchiectomy with chemotherapy and / or radiation therapy [3]. Patients can be divided into 2 groups by using Ann Arbor's staging : limited (stages I and II) and advanced tumors (stages III and IV) [6].

In limited cases; orchiectomyplays a diagnostic role in providing material for histological study and a therapeuticr ole as well considering that blood-testis barrier reduces the effectiveness of systemic chemotherapy [1]. Currently R-CHOP (rituximab, Cysclophosphamide, doxorubicin, vincristine and prednisolone) have showed good results on overall survival and progression-free survival [11]. In addition, due to the high risk of extension to the central nervous system, some authors recommend prophylactic intrathecal chemotherapy [1]. Radiation therapy may be indicated for prophylaxis in the testis and regional lymphnodes but also in the event of invasion of the retroperitoneal lymphnodes [1].

In advanced cases, patients should be treated according to the recommendations of diffuse large B-cell lymphoma of lymphnodes. The treatment is based on chemotherapy containing anthracycline associated with rituximab , scrotal radiotherapy and intrathecal chemotherapy [1].

There is no standard treatment for relapse of DLBCL. But often it is treated as a non-Hodgkin aggressive lymphoma [1]. Therapeutic decision is influenced by age, general medical state and treatments already administered [1-6].

The risk of recurrence is high even in stage I and II and mayoften occur with in 2 years[6]. The main factors of poor prognosis recognized are: age>65 years, a significant tumor mass (> 10 cm), stages III and IV of Ann Arbor staging, signs of tumor agressivity, a high level of LDH and local invasion of the epididymis and spermaticcord [12]. Also, the International Prognostic Index and its components have been frequently reported as prognostic factors including in addition to pre-cited items: advanced stage, B symptoms, ECOG performance status, Raised serum b2-microglobulin, hypoalbuminemia, involvement of the left test is[13].

Conclusions

Diffuse large B cell Lymphoma is a rare entity. A careful tumor staging is required to confirm its primitive origin. Through this case report, we have shown the importance of anamnes is for diagnosing testicular lymphoma and opting for a conservative treatment to preserve the testis in a monorchid.

Declarations

Availability of data and material:

All data and materials are available and reproductible.

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Ethics Commission

The Ethics Committee of the Robert Boulin Hospital approved the completion of ourstudy.

Consent to publish

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of the journal.

Abbreviations

R-C HOP: Rituximab, Cysclophosphamide, Doxorubicin, Vincristine and Prednisolone.

CT: Computed tomography

DLBCL: Diffuse large B-cell lymphoma

Competing interests

The authors declare that they have no competing interests.

Authors' contributions

YK was the principal author and major contribut or in writing the manuscript. TS, TG, DA, FT, MHF and FJ analyzed and interpreted the patient data and reviewed the literature. All authors read and approved the final manuscript.

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