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CASE REPORT

Primary T-Cell Lymphoma of the Testis Presenting in a Young Adult

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ABSTRACT
We report the case of a 40-year-old male presented with a painless right testicular swelling. Right radical orchidectomy was performed. The pathological diagnosis was peripheral T-Cell lymphoma-not otherwise specified (PTCL-NOS). According to Ann Arbor staging, the initial clinical stage was IEa. Treating him with four courses of the CHOEP protocol and intrathecal prophylactic chemotherapy was unsuccessful; with the appearance of orbital infiltration and a loco-regional extension. Although the patient started a second line chemotherapy, he unfortunately succumbed to death.

KEYWORDS: Testicular lymphoma; PTCL-NOS; young adult.

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INTRUDUCTION

Primary testicular lymphoma (PTL) is an uncommon and extremely aggressive lymphoma with only a few cases reported in literature [1]. The prognosis of this neoplasm is very poor and it is considered as a potentially fatal disease. Testicular lymphomas represent 5% of testicular tumors and account for approximately 1 to 2 % of all non-Hodgkin's lymphomas [2]. Diffuse large B-cell lymphoma (DLBCL) is the most frequent histological subtype, comprising more than 70% of all testicular lymphomas. PTL occurs mostly in patients of the seventh decade with an aggressive clinical presentation [3].

To the best of our knowledge, only about 21 cases have been reported in the English literature to date [4]. This lymphoma mainly occurred in Asians and Native American populations of Mexico, Central and South America [5-6]. Most PTL case reports are about primary testicular Natural Killer (NK)/T-cell lymphoma. In this manuscript we report the first case in Morocco of Primary Testicular NOS Lymphoma in a young patient and review relevant literature, to highlight the possible occurrence of this very rare and aggressive lymphoma in young adults.

CASE REPORT

A 40-year-old Moroccan man has been referred to our haematology team for management of a PTL, with a history of painless right scrotal swelling during the past two months. He had no history of trauma and no systemic B symptoms. Orchiectomy was performed as a diagnostic and initial therapy.

Ultrasonography of the scrotum demonstrated a global enlarged heterogeneous right testicle and a focal testicular mass with an increased blood flow. Computed tomography demonstrated the tissue mass within the right testicle.

A complete biological assessment including renal and liver functions, serum electrolytes, serum lactate dehydrogenase (LDH) levels were normal. Tumor markers such as alpha-fetoprotein (αFP) and beta human chorionic gonadotrophin (βHCG) were not initially performed.

The orchiectomy specimen weighted 640 g and measured 8.5 x 7 x 6 cm, attached to spermatic cord of 9 cm. The testicular section revealed a limited testicular mass measuring 7 cm of the largest diameter. Microscopic examination of the mass showed a diffuse infiltration by large lymphoid cells.

They have a scant cytoplasm and a central nucleus with granular chromatin. The spermatic cord, the epididymis were tumor free. The immunostaining showed positivity for intracytoplasmic CD3 and CD43, in association with positive immune-reactivity to CD56, granzyme B and T cell intracellular antigen-1. Pan B (CD20), CD 4, CD 5, CD 7 and CD 8 were negative. The pathological diagnosis was peripheral T-Cell lymphoma-not otherwise specified (PTCL-NOS).

The patient underwent a full staging workup including computed tomography of the chest, abdomen, pelvis and nasopharynx, bone marrow biopsy and cytology of the cerebrospinal fluid. None of which showed any other extra-testicular involvement or lymphnodes. The diagnosis of stage IEa PTCL-NOS was made.

The therapeutic decision of the multidisciplinary team discussion consisted of systemic chemotherapy with 4 courses of the CHOEP regimen (cyclophosphamide, doxorubicin, vincristine, Etoposide and prednisone) with intra-thecal Methotrexate followed up after remission of autologous stem cell transplantation.

The patient received the first-line chemotherapy with 4 courses of CHOEP regimen. Two weeks after the fourth course, the patient complained of a progressively increasing swelling of the inner corner of the left eye. He was initially treated symptomatically (antibiotics and steroids) by the ophthalmologist. The orbital swelling was rapidly progressive with installation of diplopia, binocular vision and nasal voice. Orbital MRI was then requested, which demonstrated a left pre and retro septal intra-orbital process with extension to the nasal fossa, nasopharynx and to the homolateral maxillary and sphenoid sinuses.

Upon admission to hospital, physical examination revealed in addition to the left orbital tumor several nodules with an ill-defined margin on the skin of the arms. After the performance of a left orbital biopsy, the patient was started on chemotherapy with cycle A and B of Hyper CVAD protocol (Cyclophosphamide, Vincristine, Doxorubicin, Dexamethasone /Methotrexate and Cytarabine). The evolution was complicated by the occurrence of febrile neutropenia. Unfortunately the patient succumbed to death due to a sepsis.

DISCUSSION

Primary testicular lymphoma (PTL) is an uncommon disease accounting for 1% of non-Hodgkin's lymphoma. The most common histological type of primary testicular lymphoma is diffuse large B-cell lymphoma [7].

In this paper we report a case of a young adult with a localized lymphoma of the testis. To the best of our knowledge, this is the first reported case of confirmed T cell lymphoma type NOS arising from the testis. So far, only a few reports of NK/T cell lymphoma have been described in literature. PTL is an entity that present as advanced disease, with an aggressive behavior, widespread dissemination and very poor prognosis [8].

The most common clinical manifestation of this entity is a unilateral painless testicular swelling, with a frequent vascular invasion, epididymis and cord involvement [9].

The disease tends to relapse promptly after initial management involving orchiectomy and chemo-radiotherapy [10]. Unfortunately most subjects with stage I and II lymphoma will experience relapse, which mainly occurs within the first two years of follow up [11]. Prognosis of this neoplasm is often poor even in the localized disease. Indeed, despite an aggressive treatment regimen, our patient relapses in therapy.

This lymphoma is characterized by his rare occurrence. In fact, from 1993 to 2018, only about 21 cases of NK/T cell lymphoma were reported, none of T NOS cell lymphoma. PTL is particularly frequent in china. Age of patients included in pervious repoted cases ranges from 28 to 71 years with a median age of 49.5 years. Almost all cases of literature even younger patients and those with localized diseases in the testis still have a short survival time not exceeding 12 months.

Due to the rarity of this neoplasm, optimal management is yet to be established. The treatment usually includes the performance of an initial diagnostic and therapeutic orchidectomy. Adjuvant chemotherapy or scrotal radiation or a combination is often advocated to optimize tumor control and survival [11].

This case report and literature review show that the natural history of primary T cell lymphoma of the testis presents unique features which deserve special attention to make the appropriate diagnosis and the optimal management.

CONCLUSION

We would like to share this particular case with the medical community to enrich the literature and highlight the rarity and the severity of its prognosis whatever the treatment proposed. Through this case we hope to participate to a better recognition and management of this entity. The publication of more cases of testicular lymphoma is needed to establish more appropriate protocols to improve survival and quality of life for these patients.

COMPETING INTERESTS

The authors declare no competing interest

AUTHORS' CONTRIBUTIONS.

Koubila Nisrine: has played a substantial role in designing the article

Bendari Mounia, being the principal and corresponding author, has played an important role in redaction and literature searching

Matrane Wafaa: co-author, have carried out a revision of the text

Qachouh mereym and Quessar asmaa: have taken equal parts in drafting the article.

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