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Case Report

Unilateral Primary Testicular Diffuse Large B-cell Lymphoma - A Rare Case

S A Deshpande¹, S V Suvernakar², R.D. Hanumante³, Vikas Dagar⁴, Ashish Shinde⁴, Apoorva Bhure⁴

¹Professor and Head, ²Associate Professor, ³Assistant Professor, ⁴Junior Resident, Dept. of Pathology, Dr SCGMC Nanded, Maharashtra.

Corresponding Author: S A Deshpande

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ABSTRACT

Testicular lymphoma was first reported by Malassez and Curling in 1866. Primary testicular lymphoma constitutes only 1-7% of all testicular neoplasms and less than 1% of all non Hodgkin lymphoma. We report the case of a 65-year-old man without a particular past medical history, who presented with a painless Right testicular swelling that he has noticed for 6 to 8 months. Radiological findings consisted in large heterogenous lesion with multiple tiny calcified masses that corresponded in histological examination to a diffuse intratubular lymphomatous infiltration involving spermatic cord. Immunohistochemical study showed positivity of B-cell marker (CD20) leukocyte common antigen and oct 34, placental alkaline phosphatase, CD 30 and CD 3 were negative. Ki-67 proliferation index was 70% of neoplastic lymphoid infiltration. The diagnosis of primary diffuse large B-cell lymphoma of testis was made. The patient is now treated by chemotherapy. Primary testicular lymphoma is a rare tumor whose diagnosis is based on histological findings. There are non consensual etiological or predisposing factors. Treatment modalities consist in surgical excision, chemotherapy and radiation therapy but the accurate procedures are not standardized. Factors that have been linked to more favorable outcomes include younger patient age, localized disease, presence of sclerosis at pathologic analysis, smaller tumor size, lower histological tumor grade and lack of epididymal or spermatic cord involvement.

Keywords: Testicular lymphoma; B-cell

INTRODUCTION

Primary testicular lymphoma is a rare tumor accounting for 1% of all testicular non Hodgkin lymphoma. [1] It is defined by the primary localization of the tumour in the testis at presentation. It accounts for approximately 1% of non-Hodgkin's lymphoma, 4% of all extranodal non-Hodgkin's lymphoma and 5% of all testicular malignancies with an estimated in-

cidence of 0.26/100,000 per year. Primary testicular lymphoma (PTL) is essentially an intermediate or high-grade lymphoma, and the diffuse large-cell type is the most common. [5]

CASE REPORT

We report the case of a 65-year old man without a particular past medical history, who presented with a painful right

testicular swelling that he has noticed for 6 to 8 months. There was no reported history of trauma, night sweets, fever or chills. Scrotal examination revealed a firm and enlarged testis of size 6x5x4 cm without any regional lymphadenopathy. The remainder ofthe clinical examination noncontributory. The ultra-sound examination showed enlarged, an heterogeneous testis with multiple tiny calcified masses on CT scan Right testicular heterogeneously enhancing mass involving spermatic cord S/O neoplastic etiology. Laboratory tests, especially the serum lactate dehydrogenase (LDH = 508 U/L) was mildly raised, the serum alpha-fetoprotein (αFP) and serum beta human chorionic gonadotropin (βHCG) levels were normal. An excision of the right testis was performed. It measured 9 x 5 x 5 cm and had an attached spermatic cord of 6 cm. The testicular section revealed firm grevish white mass completely replacing testicular tissue, string sign was negative. The light microscopy demonstrated diffuse intratubular lymphomatous infiltration involving spermatic cord. The malignant cells were large with scant cytoplasm and large hyperchromatic pleomorphic nuclei along with thin fibrous septae. Immunohistochemical showed study positivity of B-cell marker (CD20) leukocyte common antigen and oct 34, placental alkaline phosphatase, CD 30 and CD 3 were negative. Ki-67 proliferation index was 70% of neoplastic lymphoid infiltration. The patient underwent full staging for lymphoma including tomography of the chest, abdomen and pelvis, and bone marrow biopsy. None of which revealed any evidence of extra-testicular involvement by lymphoma or any lymph nodes. The diagnosis of primary diffuse large B cell lymphoma was made. The patient is now treated by chemotherapy.

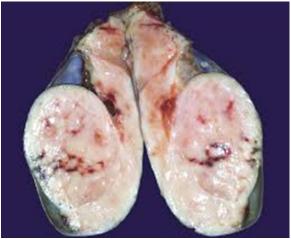


Fig 1. Gross of testis, of size 9x5x5 cm with attached spermatic cord on cut section firm, greyish white yellowish homogenous mass ,string sign negative. specimen shows complete replacement of the testicular parenchyma by fleshy,pink-tan ,yellowish tumour with involvement of the spermatic cord.

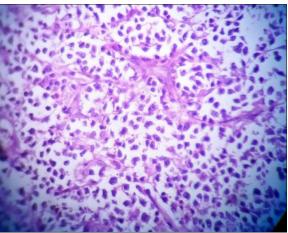


Fig 2. Hematoxylin and eosin stained section (40X) showing testicular tissue replaced by tumour cells and fibrous septae at places.

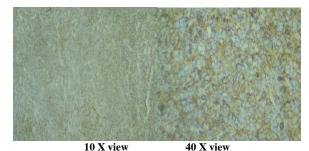


Fig 3. Showing immunohistochemistry for Leukocyte common antigen (LCA) CD 45 and labelled positive A) 10 X and 40X $\,$

DISCUSSION

Primary testicular lymphoma constitutes only 1-7% of all testicular

neoplasms and less than 1% of all non Hodgkin lymphoma. [4] The mean age at presentation is 60 years which matches with our case. The typical presentation is a testicular painless mass of variable size that usually unilateral. However, presentation, a bilateral involvement is noticed in up to 10% of the cases. [6] This fact made many authors suppose the possible multicenter origin since there is no direct lymphatic or venous connection between the right and left testis, but the fact that there are patients, like our patient, who have had localized disease and have been cured through orchidectomy alone favors the existence of testicular non Hodgkin lymphoma as primary disease. [4]

Primary testicular lymphoma has tendency to spread to several extra-nodal sites including the central nervous system (CNS), skin, lung, pleura, waldeyer's ring, soft tissue and eyes. [1,7] The imaging features reflect its infiltrative but nondestructive characteristics. At ultra-sound examination, the normal homogeneous echogenic testis is replaced focally or diffusely with hypoechoic vascular lymphomatous tissue.LDH levels have been correlated with tumour aggressiveness, whereas other tumour markers such as βHCG and αFP are rarely elevated in NHL cases. [4] In our case, the LDH was mildly raised and BHCG and aFP levels were normal.

Histological examination is the only means of diagnosis. It can be made on biopsy or surgical specimen. Testicular lymphoma is locally aggressive and can typically infiltrate the epididymis, spermatic cord or scrotal skin. [8] The predominant histology is diffuse large B-cell lymphoma (DLBCL). [4] It is reported in more than 70% of the cases. [6] The other sub-types include follicular lymphoma, plasmacytoma, lymphoblastic and Burkitt's like lymphoma. The DLBCL is classified as germinal center

B-cell-like or non germinal center B-cell-like by means of immunohistochemical expression of CD10, bcl 6 and MUM1. [9] The non-germinal center B-cell-like subgroup is the most frequent; it exhibits a high proliferative activity. [4] On the other hand, the germinal center B-cell type, like our reported case, is seen mostly in HIV-positive patients and has a better overall survival.

Histopathological differentiation of testicular lymphomas from germinal tumors is usually a challenge but these lymphomas generally appear more lobulated with well defined borders at ultra-sound examination. Other conditions might mimic testicular lymphoma such as granulomatous orchitis, palsmacytoma pseudolymphoma, rhabdomyosarcoma. There are consensual etiological or predisposing factors. Various reports have implicated prior trauma, chronic orchitis, cryptorchidism and filariasis of spermatic cord as risk factors. [8] Treatment modalities consist in surgical excision, chemotherapy and radiation therapy, but the accurate procedures are not standardized. Early retrospective studies indicated that local treatment with surgery alone or surgery plus radiotherapy and chemotherapy without anthracycline provides suboptimal disease control even in localized disease. Recently, combined modality treatment with systemic doxorubicine-based chemotherapy, prophylactic intrathecal chemotherapy and scrotal radiotherapy has been recommended because of the relapse risk at extra-nodal sites such as the CNS and contralateral testis. Despite these more aggressive treatment modalities, prognosis is often poor, even in the localized disease with the two-year relapse rate exceeding 50%. Factors that have been linked to more favorable outcomes include younger patient age, localized disease, presence of sclerosis at pathologic analysis, smaller tumour size,

lower histological tumour grade and lack of epididymal or spermatic cord involvement.

[8] According to these prognostic factors, our patient seems to have a Bad outcome because of his old age, high proliferative index.

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