A Rare Case of Primary Diffuse Large Cell Non Hodgkin's Lymphoma of Testis

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Abstract
Primary testicular lymphomas (PTL) are rare entities representing 1-2% of Non-Hodgkin's Lymphoma (NHL) and 1-7% of malignant testicular tumours. They are most commonly seen in men older than 60 years. The most common type of primary testicular lymphoma (PTL) is diffuse large B-cell type which has the potential for aggressive clinical behaviour. We report the case of a 45 years old male with primary testicular lymphoma which was first diagnosed on Fine Needle Aspiration Cytology (FNAC) and subsequently confirmed histopathologically and immunophenotypically as Diffuse Large B-Cell Lymphoma (DLBCL) showing CD20 positivity. This case report confirms the literature data concerning the rarity of testicular Non Hodgkin's Lymphoma (TNHL) with DLBCL being the most common phenotype and the poor prognosis of the disease despite aggressive management.

Key Words
Primary Testicular Lymphoma, Diffuse large cell lymphoma, Immunohistochemistry

Introduction
Primary Testicular Lymphoma is a rare entity with an incidence of only 1-7% of all testicular neoplasms (1,3,5). It is the most common testicular tumour in elderly persons with an average age of 60-80 years, but it can also occur in children (2-5). Immunophenotypically, most of these tumours are B-cell type with diffuse large B-cell subtype being the most common (1-9). It is a unique tumour because of its rarity and poses a diagnostic and therapeutic challenge.

Case Report
45 years old man presented with two months history of painless right testicular swelling which was progressively increasing in size. There was no history of trauma, night sweats, fever or chills. Scrotal examination revealed a firm and enlarged right testis. Examination of the left testis was unremarkable. There was no generalised lymphadenopathy on general physical examination. The liver and spleen were of normal size. Fine needle aspiration cytology was performed which showed uniform cell population of large lymphoid cells having round vesicular nuclei and scanty cytoplasm (Fig. 1). A cytodiagnosis of Non Hodgkin Lymphoma - Diffuse large cell type was suggested. Right orchidectomy was done. Grossly, the right testis measured 7× 5× 2 cm. On cut section, a fleshy white to tan homogenous mass replacing whole of the testis was identified. No areas of haemorrhage or necrosis were found (Fig. 2 A). Histopathologically, there was diffuse infiltration of the parenchyma by neoplastic lymphoid cells. The malignant cells were large with scant amount of pale cytoplasm.
vesicular nuclei and distinct nucleoli (Fig. 2B). Abnormal mitosis was also appreciated. Spermatic cord and epididymis showed no infiltration by the tumor cells. Immunohistochemically, tumour cells showed positivity for CD20 (Fig. 3). Based on these morphological and immunohistochemical features a diagnosis of testicular Non-Hodgkin's Lymphoma-Diffuse Large B-Cell type was made. Cerebrospinal fluid examination and bone biopsy done for staging did not show any tumour infiltration. Patient underwent radiotherapy, but was lost to treatment in between. However, after researching him one year later, it was found that patient died six months after the diagnosis.

Discussion

Primary Testicular Non Hodgkin Lymphoma was first described as a clinical entity by Malassez and Curling in 1866 (1,3,4). Since then, it has attracted attention because of its rarity and poor prognosis (3,4). Although PTL is the most common form of testicular cancer in elderly and accounting for 1-7% of all testicular tumours, it carries only 1% incidence of all Non Hodgkin Lymphomas (1-5). The mean age at presentation is 60 years, however more recent reports have indicated its incidence in younger age group also (1,6). And our patient unusually presented at 45 years of age. The typical presentation is a painless testicular mass that is usually unilateral (1,6). However, bilateral involvement can occur at presentation in upto 18% cases (1,3,6). Bilaterality supports the multicentric origin of the tumour since there is no direct lymphatic or venous connection between the right and left testis (1,3). Patients who have localised disease and have been cured through orchidectomy alone favours the existence of Testicular Non Hodgkin Lymphoma as primary disease (1,3). Although TNHLs encompass a heterogeneous group of lymphomas, the most common PTL is diffuse large B-cell lymphoma- intermediate grade which comprises 65-70% of the cases (1,2,3,6). The other subtypes that are frequently reported include Follicular...
Lymphoma, Lymphoblastic and Burkitt’s like Lymphoma (1,6). Diffuse large B-cell lymphoma is classified as germinal center B-cell like and non-germinal center B-cell like by means of immunohistochemical expression of CD10, Bcl-6 and MUM1 (1,3,6). The majority TNHL cases belong to non germinal center B-cell like subgroup (1,3,6).

Lymphomas are one of a triad of tumours along with Spermocytic Seminoma and Metastatic tumours which pathologists should particularly consider, assuming reasonably appropriate morphology in patients older than 60 years (5). But the more lobulated mass with well defined borders on ultrasonography decreases the challenge (1,5). Moreover, it is locally aggressive tumour with more often infiltration into epididymis, spermatic cord or scrotal skin which the other entities rarely show (1,5). Other conditions that may mimic testicular lymphomas include Granulomatous Orchitis, Pseudolymphoma, Plasmacytoma and Rhabdomyosarcoma (1,3,5). PTL shows a propensity for extra nodal metastasis which may be seen at the time of presentation or develop during the course of the disease. Common extranodal sites are contralateral testis, central nervous system (CNS), skin, lung, pleura, Waldeyer’s ring, eyes and soft tissues (1,3,5,6,7). There are no well documented etiological or predisposing factors or any significant associations between history of trauma, chronic orchitis or cryptorchidism and subsequent development of TNHL (3). The management of patients with TNHL presents several challenges (2). An aggressive treatment approach is required in the form of orchidectomy, systemic chemotherapy, prophylactic intrathecal chemotherapy and scrotal radiotherapy (1). Despite the recent advanced and aggressive treatment, the prognosis is often poor (1). Factors that show good prognosis include younger age, unilateral localised disease, presence of sclerosis, small tumour size, lower histological tumour grade and lack of epididymal or spermatic cord involvement (1,5,7).

Conclusion
Although testicular lymphoma was identified more than 100 years ago, it remains a subset of interest since it follows a unique biological and clinical course and optimal treatment continues to be controversial. The treatment will continue to evolve with improved understanding of the molecular and genetic characteristics of testicular lymphoma, identification of patients at higher risk of relapse and with incorporation of newer drugs into current regimes of chemotherapy.

References