

A rare case of primary extra-nodal large cell Non-Hodgkin's Lymphoma of testis

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Citation

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Abstract

Non-Hodgkin lymphoma (NHL) of the testes is an uncommon extranodal presentation with an incidence rate of 0.06 to 0.09 per 100,000 persons and accounts for about 1% of all NHLs and 5% of all testicular tumors. The disease typically presents in patients aged over 60 years and is the most common testicular tumor in patients older than 60 years. Right- and left-sided testicular involvement is equal in frequency and approximately 6% of testicular lymphomas will have bilateral involvement. Testicular NHL has a relapse site in the central nervous system (CNS). It is actually a rare disease. Our case showed typical features of NHL without any CNS involvement even after 5 years. The patient was offered a standard chemotherapy regimen without any radiotherapy.

CASE HISTORY

A 50-year old male patient residing in Mumbai, fisherman by occupation, was admitted with complaints of right-sided painless scrotal swelling for 3 months which was progressively increasing in size. The patient had no history of trauma, fever with chills or loss of testicular sensations. He did not take any specific treatment. He came to our institute for heaviness in the right half of his scrotum and did not have any major medical illnesses nor had he undergone any surgery in the past.

The patient was chronic tobacco chewer and alcoholic.

On examination, his vital and systematic parameters were within normal limits (WNL). There was no systemic abnormality. There was no lymphadenopathy, either.

On local examination, his left hemi-scrotum was WNL. His right hemi-scrotum had a large solitary non-tender swelling measuring 10 × 6 × 4cm, hard in consistency; his temperature was WNL. Testicular sensations were maintained. The swelling was non-fluctuant and non-transilluminant.²

Inguinal lymph nodes were not palpable. Per rectal examination was WNL.

The patient was investigated. USG of abdomen & pelvis did not show any lymphadenopathy. His chest x-ray was normal.

A decision to perform right inguinal orchidectomy was

taken. Under spinal anesthesia, right inguinal orchidectomy was done with 'non-touch technique'.

Gross pathology showed a thin rim of compressed testis and a large growth arising from the right testis, which was well encapsulated. The growth was yellowish-white in color. The external surface showed areas of congestion and necrosis. The cut surfaces revealed uniform homogeneous material.

Histopathology revealed a large-cell Non-Hodgkin's lymphoma.

Thereafter, a CT scan of abdomen & pelvis was done to rule out metastasis and to assess the status of liver and spleen. The CT scan did not show any spread. The patient was offered an anthracycline-based chemotherapy regimen without any radiotherapy, as the patient was lost to treatment in between. However, after meticulous follow-up after 5 years, we found the patient and repeated necessary investigations showed that the patient is healthy and asymptomatic without any evidence of recurrence, local or in the CNS.

DISCUSSION

Primary testicular lymphoma is predominantly a disease of the elderly (1). The median age in our study was 55 years compared to ≥70 years reported in other recent reports. Although multiple lymphoma classifications have been used in the description of testicular lymphoma, the following one

is most commonly accepted worldwide along with histological variants. Grading is done as low-grade, intermediate grade and high grade. Prognosis is worst in high-grade tumors. Testicular lymphomas are chiefly of 2 types: childhood lymphomas and adult variety.

Staging of NHL (ANN ARBOR CLASSIFICATION)

Stage I – Involvement of single node or extra-nodal organ/site.

Stage II – Involvement of 2 or more lymph-node regions on same side of diaphragm alone or with involvement of limited contiguous extra-lymphatic organ/tissue.

Stage III – Invasion of nodes on both sides of diaphragm, which may include spleen and/or limited contiguous extra-lymphatic organ or site.

Stage IV – Disseminated foci of one or more extra-lymphatic organs with or without nodes.

Predominant histology is diffuse large B-cell lymphoma (DLBCL). In the largest series involving 3669 cases of testicular NHL registered with the National Cancer Data Base (NCDB) from 1985 to 2004 (2), DLBCL accounted for 77.8%. Other B-cell types and T-cell types were seen in 21.1% and 1.1% of cases, respectively. In a recent series of 18 cases, immunohistochemical subtyping of testicular DLBC was attempted and it was found that the majority (89%) had the non-germinal center subgroup (CD 10/Bcl-6 negative and MUM-1 positive) with high proliferative activity.

Treatment for testicular NHL includes removal of tumor. The stage II & I patients can be given abdominal & pelvic irradiation. Stage I patients after removal of tumor should be subjected to a chemotherapy regime of cisplatin, vincristine & cyclophosphamide. Bone marrow suppression should be watched for patients under treatment. This was a typical protocol of treatment in the past.

However, in reality, the management of patients with testicular lymphoma presents several challenges (3). Because of the poor prognosis, an aggressive treatment approach is required. However, testicular lymphoma is predominantly a disease of older men who often have limited ability to tolerate aggressive treatment. Nowadays, an anthracycline-based chemotherapy regimen is offered predominantly to these patients with good response (4). There is no definitive data for use of monoclonal antibodies

(rituximab) in this subset of patients, although there are occasional case reports. Even prior to the introduction of rituximab, survival and prognosis has improved over a period of time, perhaps due to use of multimodality therapy (5). High rates of central nervous system relapses in historical series have led to a recommendation for routine CNS prophylaxis with at least intrathecal methotrexate (6). But its role in prophylaxis remains controversial as CNS relapses have been observed after intrathecal therapy and CNS relapses also occur in brain parenchyma.

To conclude, taking into account the rarity of this disease, it will be difficult to standardize the therapeutic and preventive strategies through randomized trials; 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 (7).

Our patient presented with typical diffuse large B-cell NHL of testis. He had no systemic involvement. He underwent standard high inguinal orchidectomy using non-touch technique. Postoperatively he was offered a standard cisplatin, vincristine and cyclophosphamide chemotherapy regimen. He could not receive radiotherapy, as he was lost to treatment. However, after researching him five years later, he is asymptomatic and healthy without any local or CNS recurrence.

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