Case Report

Primary T-cell Non-Hodgkin's Lymphoma Presenting as a case of Testicular Mass

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Abstract

Introduction: T-cell lymphoma is a very rare tumor. Only a few cases have been reported in world literature. It involves the testis infrequently, which deserves special attention because of the poor prognosis and the need to make an appropriate diagnosis, which can lead to a better therapeutic strategy.

Case presentation: A 34-year-old man presented with left testicular swelling for past four months. The swelling was painless, hard and rubbery. Testicular ultrasound showed diffuse increase in size of the testicle, with alteration in its echogenicity. FNAC was done and the diagnosis was Non-Hodgkin's lymphomas. The patient underwent orchidectomy and basing on histopathological and immunohisto-chemical tests, a peripheral T-cell lymphoma, not otherwise specified was diagnosed.

Conclusion: Testicular peripheral T-cell lymphomas are rare and aggressive cancers which is clinically different from seminoma and other non-neoplastic conditions.

Key-words: Testicular T-cell lymphoma, Extranodal lymphoma.

Introduction

Malignant lymphomas comprise 5% of all testicular malignancies. It is the most common testicular tumor in elderly persons but it can occur in any age group including children. It has much greater tendency for bilateralism than germ cell tumors; as a matter of fact, about 50% of cases of bilateral testicular tumors are malignant lymphomas[1]. The testicular lymphomas are usually disseminated at the time of presentation[2,3]. Nearly all these cases are of non-Hodgkin's type. The phenotype of the lymphomas is almost always B-cell type with most of the tumors being the diffuse large B-cell subtype; however, rarely, anaplastic lymphoma, Burkitt's lymphoma or Hodgkin's lymphoma may involve the testis as the primary site[2,3,4]. T-cell lymphoma of the testis is rare whether as a primary or secondary tumor. Better prognosis is predicted if the tumor is a primary, localized to the testis and unilateral (5-year survival 60% vs. 17% for disseminated disease/other stages). Best treatment options are orchidectomy and chemotherapy along with radiation[5]. Recently, combined modality treatment with systemic doxorubicin-based chemotherapy, prophylactic intrathecal chemotherapy and scrotal radiation therapy has been recommended because of the relapse risk to extranodal sites such as the central nervous system and contralateral testis. Despite these, more aggressive treatment modalities, prognosis is often poor, even in the localized disease with the two-year relapse[6,7] rate exceeding 50%.

Case Presentation

A 34-year old male patient reported at the department of Pathology, Rajshahi Medical College for the purpose of FNAC in February 2014 with the history of a rapidly enlarging painless lump in his left scrotum for the past four months. There was no reported history of trauma or night sweats but he had fever for a week. There were no co-morbid conditions. His previous medical and surgical history was unremarkable. On physical examination, a non-tender lump measuring 08 × 06 cm, firm-to-hard in consistency was found as a left testicular swelling. It was associated with slight skin puckering. Examination of the other, that is, the right testis

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revealed that it was mildly enlarged. There was no generalized lymphadenopathy on general physical examination. The liver, spleen and intra abdominal lymph nodes were of normal size. Sonographic examination of the scrotum revealed a well-defined, hypoechoic space-occupying lesion in the cranial and lateral aspect of the left testis and an ill defined hypoechoic space-occupying lesion in right testis. Findings on sonographic and Computerized Tomography (CT) examinations of the abdomen were normal.

Differential diagnosis for this sonographic finding was seminoma and orchitis. His Erythrocyte Sedimentation Rate (ESR) was within normal range and the Montoux test for Mycobacterium tuberculosis was negative. His complete blood counts at the time of presentation were within normal limits. On discussion with the patient, he was informed about the pathology that would be isolated tuberculous orchitis and confirmed consent was taken for FNAC test.

FNAC test of a left testicular swelling was performed and diagnosed as a case of non-Hodgkin's lymphomas, not otherwise specified (Fig-1). Testicular biopsy was performed under general anesthesia and the specimen was sent in formalin to the department of Pathology, Rajshahi Medical College, Rajshahi. The gross morphology of the mass appeared as white tan-pink colored fleshy mass resembling a seminoma, Histologic examination of the specimen revealed a diffuse proliferation of malignant round cells within the interstitium of the testicular parenchyma and areas of necrotic foci.

The cells were present in the form of sheets with scant cytoplasm and slight variation in size and shape of medium-to-large nuclei, which frequently showed irregular folding and granular chromatin. Mitotic activity was also observed with mitoses seen at a rate of 15/10 high power fields. Intervening scattered vessels were also present (Fig-2).

Immunohistochemical stains were applied and tumor cells showed positivity with T-cell markers CD3 and CD 43 (Fig-3 & 4.). Pan B (CD20), CD 4, CD 8, CD 5 and the epithelial marker, cytokeratin was negative. The study was not experimental and no identifiable material was used.

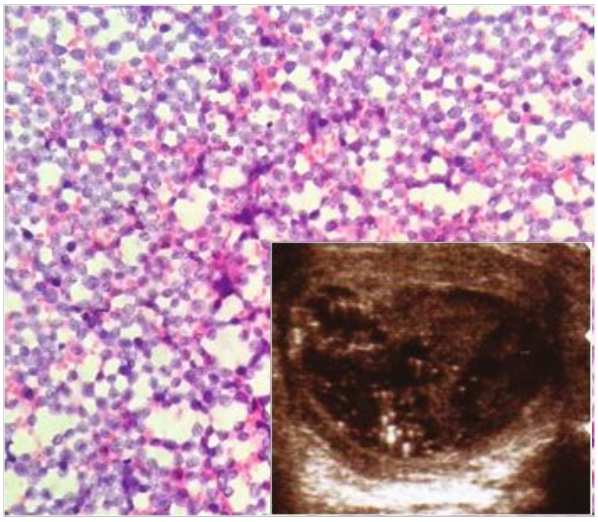

Fig-1: FNAC Test show monotonous population of immature lymphoid cells. (Inset) well-defined, hypoechoic space-occupying lesion in the cranial and lateral aspect of the left testis.

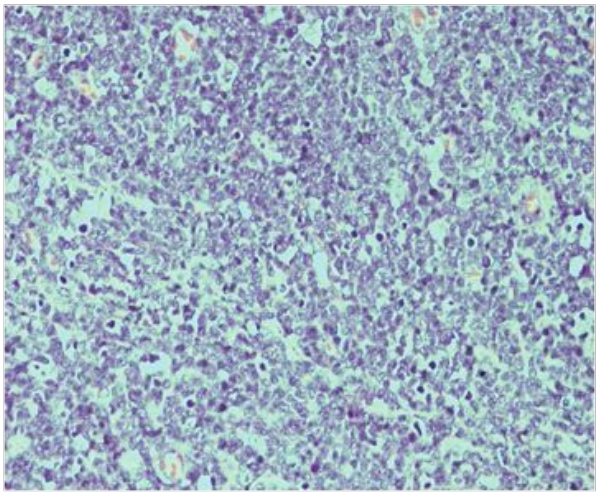

Fig-2: Atypical lymphoid cells which have narrow cytoplasm, hyperchromatic enlarged nucleus which completely destroy normal testicular structure (H&E × 400).

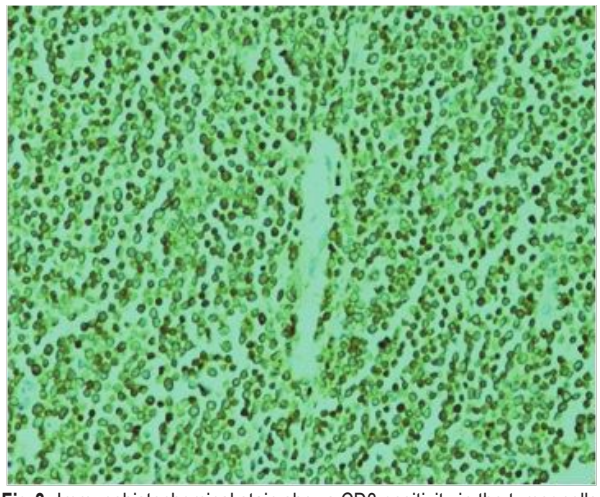

Fig-3: Immunohistochemical stain shows CD3 positivity in the tumor cells.

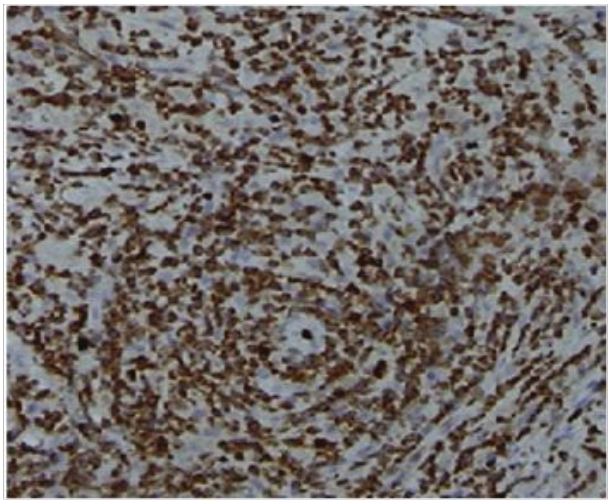

Fig-4: Immunohistochemical stain showing CD 43 was also positive in the tumor cells.

Discussion

Testicular lymphoma was first reported by Malassez and Curling[8,9] in 1866. Primary testicular lymphoma constitutes only 1-7% of all testicular neoplasms and less than 1% of all non Hodgkin's lymphoma. The mean age at presentation was 60 years, but the recent published cases concerned patients were found younger than the past are reported and considered that this fact had a positive effect on the outcome of the patients[10]. According to the recent publication, this patient is young and only 34 years old. The typical presentation is a testicular painless mass of variable size that is usually unilateral. However, while in presentation, a bilateral involvement is noticed in up to 10% of the cases[11]. Constitutional symptoms such as fever, weight loss, anorexia, night sweating and fatigue are seen in 25 to 40% of the patients[9,10].

In primary T-cell lymphomas, most reported cases are of T-cell/Natural Killer (NK) cell lymphoma with very few cases of peripheral T-cell lymphoma (NOS). Our case represents one of those few cases of peripheral T-cell lymphomas[3,4,5]. EBV-associated T-cell/NK cell lymphomas most commonly involve the nasal cavities and these are aggressive extranodal lymphomas. These are rarely encountered at sites other than the upper aero-digestive tract[12]. Clinical presentation is usually of a painless, unilateral testicular swelling. In some cases, abdominal pain with ascites being reported due to large retroperitoneal lymph nodes. This patient also had a painless testicular mass for the past four months. Associated B symptoms (fever, night sweats, weight loss) usually present only in advanced stages, accounting for 25% to 41% of patients at diagnosis and these were also not present in our patient so prominently[12,13]. A malignant lymphoma in which the tumor mass is limited to the testis at the time of clinical onset of the disease is rare. Since the first report of non-Hodgkin lymphoma manifesting as a testicular mass in 1866, primary testicular lymphoma has attracted attention because of its rarity and poor prognosis. 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[14]. 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[14,15]. LDH levels have been correlated with tumor aggressiveness, whereas other tumor markers such as βHCG and AFP are rarely elevated in TNHL cases[16].

Testicular lymphoma is locally aggressive and can typically infiltrate the epididymis, spermatic cord or scrotal skin. Histopathological differentiation of testicular lymphomas from germ cell tumors are 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, pseudo-lymphoma, and rhabdomyo-sarcoma[16]. Testicular lymphoma is a lethal disease with a median survival of approximately 12 to 24 months[3,5,13]. Our patient has no lymphadenopathy or splenohepatomegaly as demonstrated by CT. Until now, 4-monthly follow up are being done and the patient is alive. He received chemotherapy, the conventional cyclophosphamide, adriamycin, vincristine, and prednisolone (CHOP) regimen initially. However, recurrence of the mass developed after 3 months and the patient is now under radiotherapy treatment[12,13]. Testicular T-cell lymphoma deserves to be distinguished from the other testicular lymphomas for example, diffuse large B-cell lymphoma and Hodgkin's lymphoma because of the different treatment options. In fact, this lymphoma has a tendency to occur at a younger age, to disseminate early, to have an aggressive course and is strongly associated with EBV[5,17].

Conclusion

Testicular peripheral T-cell lymphomas are rare and highly aggressive cancers, with clinical and histological differentials of seminoma and non-neoplastic conditions. Correct diagnosis with immunohistochemical techniques is mandatory for the proper treatment and further directed management of these tumors.

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