Primary Testicular Non-Hodgkins Lymphoma: A Case Report

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Abstract

Here, we report a case of 71 years old male, presenting with a left sided testicular tumor without any reported inguinal, abdominal or peripheral lymphadenopathy, diagnosed as Non-Hodgkins lymphoma of testis on histo-pathological examination and confirmed by immunohistochemistry (IHC).

Key words: Testicular tumor, primary testicular lymphoma, NHL, IHC.

INTRODUCTION

Primary testicular lymphoma is an uncommon, extranodal and aggressive form of lymphoma. It accounts for 1-2% of all NHL, 2-3% of extranodal lymphomas and less than 5% of all testicular malignancies¹-³. The tumor can invade the contralateral testis and can rapidly spread to CNS⁴. Villa D et al have highlighted the high prevalence of B cell lymphoma in extranodal sites like testis, brain, breast, kidney, adrenal gland, nasopharynx, bone and bone marrow⁵. Bierman et al reported that age more than 60 yrs, low serum albumin and high serum lactate dehydrogenase (LDH) levels point towards lymphoid malignancy in testicular tumor⁶. A study by Jarosinska showed that this aggressive tumor had a high relapse rate despite having proper chemo and radiotherapy⁷. Other studies have concluded that 68% of primary testicular lymphoma are of intermediate grade DLBCL, followed by 30% high grade diffuse small non-cleaved (Burkitts and Burkitt like subtype)⁸.

Case Report

A 71 years old patient presented with a painless left sided testicular enlargement for two months. Patient was a follow-up case of recurrent left epididymo-orchitis (previous records not available). The physical examination revealed a non-tender left testicular mass of 7x4 cms size, without any inguinal lymphnode enlargement. USG of testis showed altered echotexture appearing hypoechoic, enlarged in size (5.0 x 6.8 x 7.7 cm APXTDXCC) and show increased vascularity on doppler with multiple internal septa in testis. Right testis measured 2.6x2.x0x3.7 cms. USG abdomen showed normal liver, kidney, spleen with no para-aortic or other lymphadenopathy. MRI of pelvis and abdomen showed a well defined smooth marginated lesion in left scrotal sac involving testicular parenchyma appearing iso to hypointense in both T1 and T2W images, measuring 6x5 cms in axial plane and 7.3x4.5 cms in sagittal plane, showing mild homogenous enhancement post contrast (Fig 1). No evidence of pelvic or inguinal lymphnodes seen. Rest of the abdominal study was normal. A radiological impression of testicular mass likely seminoma was given. Routine blood investigations were within normal limits. His LDH level was645 IU/L (Normal 128-287 IU/L), alpha feto protein level 2.1 ng/ml (N-0-15 ng/ml) and HCG levels 1.2 mIU/ml (Normal less than 5 mIU/ml).

A left sided high inguinal orchiectomy was done. Histopathology lab received already cut open orchiectomy specimen measuring 9x7x5 cm with attached spermatic cord 7.5 cm in length. External surface grey brown, capsulated. Cut surface showed a homogenous grey white solid mass with few small necrotic areas (Fig. 2).
Microscopic examination showed effaced testicular architecture with diffuse interstitial infiltration of non-cohesive cells with interspersed atrophic, thick walled seminiferous tubules (Fig 3). The cells have irregular nuclei, scanty cytoplasm and single to multiple nucleoli (Fig 4). Infiltration of wall of blood vessels and confluent necrotic areas also seen. Tumor involved epididymus. Distal end of spermatic cord was free of tumor. Periodic acid Schiff (PAS) stain was negative (Fig 5). Immunohistochemistry for CD20 showed diffuse positivity (Fig 8 test, Fig 9 control) and CD3 (Fig 6 test, Fig 7 control) staining was negative (Tests run with controls). A diagnosis of Diffuse B cell lymphoma was given.

**DISCUSSION**

Histologically, 80-90% of primary testicular lymphomas are Diffuse large B cell type6. 60% of patients in stage I, while 30% are in stage II and bilateral testicular involvement is seen in approximately 35% of cases7. Most frequent symptom is painless testicular swelling, fever, night sweats, weight loss and fatigue are also seen. Granulomatous orchitis, pseudolymphoma, plasmacytoma, seminoma, embryonal carcinoma are the differential diagnoses. Intra-tubular germ cell neoplasia, the precursor lesion of testicular tumors is not seen in primary testicular lymphoma8.

Various subtypes of primary non-hodgkins lymphoma of testis include Diffuse large B cell lymphoma, Burkits lymphoma, follicular lymphoma and Large cell anaplastic lymphoma.
Usual H&E stained sections show interstitial growth pattern with effacement of testicular architecture. Seminiferous tubules may be seen. In preserved tubules spermatogenic arrest, interstitial fibrosis and tubular hyalinization can be seen. The tumor is composed of discohesive sheets of medium to large sized lymphoid cells with round to oval vesicular nuclei having fine chromatin, scanty cytoplasm and multiple nucleoli.

Seminoma cells are uniform with abundant clear (PAS positive) cytoplasm, sharply outlined cell membranes, a large centrally located nucleus and clumped chromatin. Nucleoli are prominent. The cells are reactive for placental alkaline phosphatase (PLAP), CD117, LDH, Vimentin, ferritin but negative for HMWCK, EMA and CD30, which differentiates seminoma from embryonal carcinoma. Embryonal carcinoma shows a pattern of growth with carcinomatous appearance. The cells are more anaplastic with numerous mitoses and marked pleomorphism. Granulomatous orchitis identified by a granulomatous lesion centered in seminiferous tubules.

Advance stage tumors tend to spread to extranodal areas such as CNS, skin, waldeyers ring and lungs. Staging of primary testicular lymphoma, modified by Nordic lymphoma group is as follows: stage I: Unilateral testis involvement with or without epididymus or cord involvement; stage II: Abdominal and pelvic lymphode involvement; stage II-IV: distant metastasis.

The most important factor identifying prognosis are clinical stage and histological grade. A primary tumor more than 9 cm, presence of spermatic cord, epididymus and bilateral testis involvement, vascular invasion, high LDH levels, presence of B symptoms, high international prognosis score and left testis involvement are factors associated with poor prognosis.

The treatment for stage I and II tumors consists of orchectomy with chemotherapy (R-CHOP; rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone). For stage III-IV disease, systemic chemotherapy, scrotalradiotherapy and intrathecal chemotherapy are preferred.

CONCLUSION

Primary testicular lymphoma can be isolated in testis without manifestation of systemic disease. The contralateral testis should always be taken into consideration.

REFERENCES