

Case Report

Primary Testicular Lymphoma: A Rare Entity

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Abstract

Introduction: Primary testicular lymphoma is a rare, aggressive form of tumor and accounts for 1% of all extranodal lymphoma. It carries a poor prognosis. So an accurate early diagnosis is essential.

Presentation of case: Herein we are reporting a case of this unusual tumor in a 73 year old male patient who presented with a large right testicular swelling. Histological examination showed feature of testicular lymphoma.

Conclusion: This tumor can easily be misdiagnosed as spermatocytic seminoma, so immunohistochemistry is necessary and this rare entity should always be kept in mind among the differential list of all testicular masses.

Keywords

- Testis
- Lymphoma
- CD45

INTRODUCTION

Primary testicular lymphoma is an uncommon, extranodal and aggressive form of lymphoma. It accounts for 1% of all NHL and 5% of all testicular tumors [1]. Despite this, it is the most commonly observed testicular malignancy in men above 60 years and histopathologically 80%-90% of primary lymphoma of testis is of DLBCL type. Aim of our case report is of discussing histomorphological feature of this rare malignancy so that this differential diagnosis had to be kept in mind in case of any testicular swelling of unknown etiology.

CASE PRESENTATION

A 73 year old man presented with right sided testicular swelling gradually increasing in size for last the two months. There was no history of trauma or congenital anomaly of the testis. He denied any associated pain or fever in the meantime. On examination there was a mass in right side of the scrotum approximate 6cm x 6cm in size. It was firm in consistency having smooth surface without any tenderness. No lymphadenopathy was present at that time. USG of the inguinoscrotal region showed a hypochoic lesion at right testis with normal paratesticular tissue. The other testis was free. His routine investigation (complete hemogram, LFT, ECG, Chest X-ray) was within normal range. A provisional diagnosis of right sided malignant testicular tumor was made by the surgeons and high inguinal orchidectomy was done. Specimen of the testis along with cord was sent to our pathology department for histopathological examination (Figure 1).

Grossly, the testis measured 6cm x 5cm x 3cm. A well encapsulated mass was present and it involved almost the entire

testis. Cut surface showed grey- white homogenous region with nodularity and focal areas of necrosis. The histopathological finding revealed effaced testicular architecture with diffuse proliferation of malignant mononuclear uniform cells separated by fibrous stroma. Individual cells were relatively monomorphic, non cohesive with scanty cytoplasm. Nuclei were large, irregular with indistinct nucleoli. In few areas tumor cells were seemingly infiltrating the interstitial space around the atrophic tubules. Margin of the spermatic cord was involved and there was no feature of intratubular germ cell neoplasia. From the histomorphological feature we could not determine whether it was a germ cell neoplasia or lymphoid tumor of the testis. Immunohistochemistry was done for CD45 & PLAP and it confirmed the lymphoid nature of the tumor cell (CD45 positive) and differentiates it from its closest D/D that is spermatocytic seminoma (PLAP negative). If embryonal carcinoma comes in differential diagnosis, then we should go for c-kit and OCT4 (Figure 2).

Patient was referred to oncology department for chemotherapy and he is still under follow up.

DISCUSSION

Most commonly lymphoma involves the testis through dissemination from the extra testicular sites (secondary testicular lymphoma) but the involvement of the testis without systemic lymphoma (primary) is a rare incident.

Malassez et al., first reported a case on testicular lymphoma in the year 1897 [2]. It is a malignancy of elderly. Risk factors are immunosuppression, chronic orchitis, trauma and filarial [3]. They usually present with rapidly progressive painless enlargement of

the testis. Bilateral testicular involvement is usually metachronous in nature and occurs in approximately 20% of patients [4]. Systemic symptoms such as fever, anorexia, night sweat and weight loss may be present. There may be local spread to the epididymis, spermatic cord and scrotal skin with sharp scrotal pain [5]. Extranodal metastasis can be present at the time of diagnosis. Among them CNS is the most common site. Waldeyer's ring, urogenital tract, lung and liver are rarely involved. In our case patient had no B symptoms.

The most important factors that determine prognosis are primary tumor larger than 9 cm, involvement of epididymis and spermatic cord, bilateral involvement, vascular invasion, advanced age, presence of B symptoms and left testis involvement [6]. Of the known poor prognostic factors, our patient had advanced age and cord involvement.

Differential diagnosis of testicular lymphoma includes few non neoplastic conditions like viral and granulomatous orchitis and malignant neoplasms specially classical and spermatocytic seminoma. Heterogenous and benign appearing inflammatory cellular infiltration of orchitis, differ from lymphoma by more homogenous and malignant infiltration (Figure 3). Classical seminoma cells are uniform, have distinct cell membrane, abundant glycogen rich cytoplasm, central nuclei with prominent nucleoli (Figure 4). Spermatocytic seminomas have three distinct cell populations and it gives PLAP positivity that is not present in lymphoma.

Regarding treatment, after orchidectomy there is a high

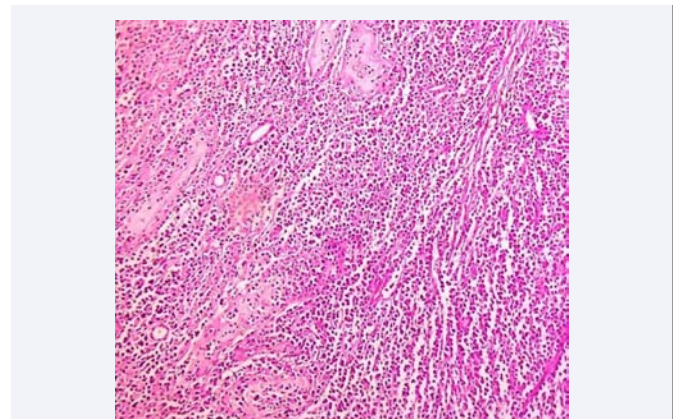


Figure 3 Diffuse infiltration of malignant lymphoid cells around the atrophic tubules (H & E stain; 10x).

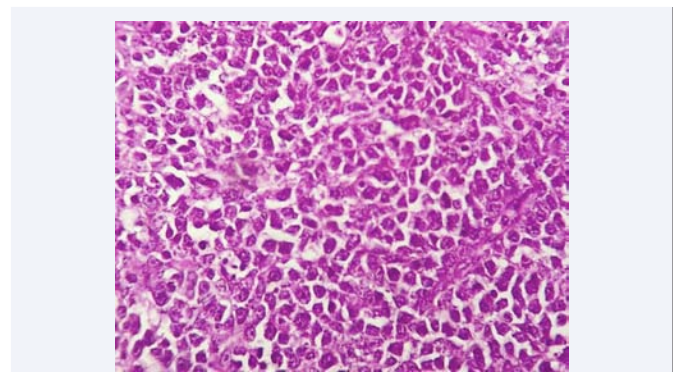


Figure 4 Individual cells are non cohesive having scanty cytoplasm, large hyperchromatic nuclei with indistinct nucleoli (H & E stain; 40 x).



Figure 1 Specimen of testis a) showing greyish white solid mass b) on cut section having nodularity and homogenous architecture.

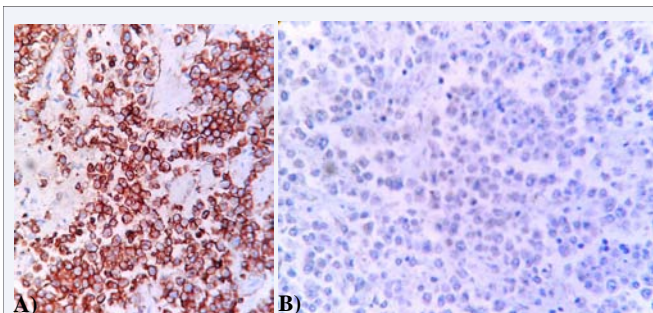


Figure 2 Immunohistochemistry showing a) CD45 positivity b) PLAP negativity.

chance of relapse in these patients. According to Danish series the median survival after relapse is < 2 months [1]. It is presumed that high frequency of relapse in CNS and contralateral testis, elderly age group and associated comorbidity comprise narrow time gap between time of diagnosis and fatal outcome. But now the scenario is changing and outcome of the patients are improving. Various clinical trials are going on regarding its therapy. As its management differ completely from germ cell neoplasm, here lies the importance of pathological examination of the specimen so that we can identify it correctly and distinguish it from other entity that would help the future treatment policy.

CONCLUSION

Primary testicular lymphoma is a disease with poor prognosis. The rare incidence of the disease, its development and tumor behaviour are different from the germ cell tumor of testis. As there is no definite therapeutic protocol after orchidectomy, it should be considered as a manifestation of systemic disease of the testis. Relapse of the CNS and involvement of the contralateral testis should be taken into consideration. For this reason, lymphoma should be kept in mind for the patient who present with a mass in the testis and surgeon, pathologist and oncologist should take a joint action.

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