Case Report

Primary DLBCL of testis: Presenting as a case of testicular mass

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ABSTRACT

Primary testicular non-Hodgkin's lymphoma (NHL) is a rare, clinically aggressive form of extranodal lymphoma and constitutes 1% of all NHL and 5% of all testicular neoplasms. The vast majority of primary testicular lymphomas (PTLs) are diffuse large B-cell lymphoma (DLBCL); however, Burkitt's lymphoma, anaplastic lymphoma, or Hodgkin's lymphoma may primarily involve the testis but it is less common. High inguinal orchiectomy, along with chemotherapy with or without radiotherapy, is the current treatment of choice for these tumors. Here, we present the case of primary testicular NHL in a 72-year-old male.

Key words: Diffuse large B-cell lymphoma, Non-Hodgkin's lymphoma, Primary testicular lymphoma

Testicular lymphoma is a deadly disease, representing 1–2% of all non-Hodgkin lymphomas (NHLs) and approximately 55% of all testicular lymphomas [1,2]. It is the most common testicular tumor in males between 60 and 80 years of age [3]. The majority of patients with this disease are categorized as having diffuse large B-cell lymphoma (DLBCL), histological subtype, accounting for 80–90% testicular lymphomas [2,4,5]. Testicular lymphomas differ from germ cell and stromal testicular tumors by their frequent bilateral testis involvement, bilateral meta-synchronized testis involvement, and synchronous involvement [6]. Primary testicular lymphomas have the worst prognosis with only a 5-year survival rate. Since these are aggressive tumors with a worse prognosis, these cases should be reported in detail for appropriate understanding and treatment of patients. These tumors should be differentiated from other scrotal masses such as germ cells tumors of the testis or other NHL and appropriate multimodality treatment should be given to the patient [7].

We present this case of primary testicular NHL in a 72-year-old male.

CASE REPORT

A 72-year-old elderly male presented with a complaint of the right testicular swelling for the past 1 month. The swelling was of gradual onset and not associated with pain. There was no history of trauma. The patient was a known case of Parkinson's disease.

General examination showed that the patient was afebrile, not anemic, or jaundiced and there was no cyanosis or clubbing or generalized lymphadenopathy but pedal edema was present. The blood pressure was 130/90 mmHg and pulse rate was 78/min. Local examination revealed swelling in the right testicular region measuring 14 × 8 cm, soft in consistency, non-tender, and no redness. The skin over the swelling did not show any ulcer or pigmentation.

All routine blood investigations were done. Complete hemogram showed hemoglobin 11.7 g/dL, hematocrit 36.2%, RBC count 3.99 million/cu.mm, platelet count 261,000/cu.mm, and total WBC count 5200/cu.mm. Urine routine examination showed pus cells 15–18/HPF, granular, and hyaline casts. Anti-HIV, HBsAg, and anti-HCV were negative. Blood sugar was 121 mg/dL, urea was 44 mg/dL, and serum creatinine was 1.07 mg/dL. Serum total beta-human chorionic gonadotropin (HCG) was below 1.2 mIU/mL and serum alpha-fetoprotein was 3.86 IU/mL. COVID-19 RT-PCR was negative. The patient was taken up for surgery after the anesthetist's consent.

The patient underwent right high orchidectomy under spinal anesthesia. The right inguinal incision was made, the inguinal canal was opened, and cord structures were identified. Two nodules were seen at the level of superficial ring and another at mid-cord structures. Cord structures were doubly ligated at the internal ring and divided. Testis was brought through the inguinal incision and orchidectomy was completed. Hemostasis was secured. The wound was repaired in layers and a 14F Foley catheter was placed. The post-operative period was uneventful.

After surgery, we received a right orchidectomy specimen measuring 13 × 8 × 7 cm. The external surface was smooth, cut surface showed a pale brown, soft to firm lesion measuring 7.5 × 7 cm with areas of hemorrhage. Spermatic cord measured

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9 cm, it was thickened and congested. Microscopic examination showed testicular tissue with an infiltrating neoplasm composed of tumor cells arranged in solid sheets (Fig. 1a). The tumor cells showed pleomorphic nuclei, irregular nuclear border, coarse chromatin, conspicuous nucleoli, and scanty cytoplasm (Fig. 1b). Frequent mitotic activity of 4–6/HPF was seen. Foci of necrosis were also noted. The tumor cell infiltrates the tunica, spermatic cord, and epididymis. The diagnosis was given as undifferentiated carcinoma and immunohistochemistry (IHC) was suggested to rule out NHL. On IHC, the tumor cells were immunopositive for CD45, CD20, BCL-2, and CD10 (patchy) while they were immunonegative for CD3, CD5, and MUM-1. BCL-6 was non-contributory (Fig. 2). Ki-67 index was 80% in the highest proliferating area. Hence, the final diagnosis was given as high-grade NHL-diffuse large B-cell type.

The patient was referred to a higher center for medical oncologist follow-up and management. Hence, the patient could not be followed up.

DISCUSSION

Lymphomas primarily involving the testis are rare and account for only 1–2% of all NHL [1]. Most commonly seen in elderly males with a median age of presentation of 60 years [1,2,6]. Since the patient presents with painless, slow-growing, and testicular enlargement, elderly males with testicular swelling should be examined thoroughly for diagnosing this rare yet aggressive malignancy. The patient may also have other symptoms such as fever, loss of appetite, loss of weight, fatigue, and night sweats. Most testicular tumors have intratubular germ cell neoplasia as a precursor lesion but this is not seen in primary testicular lymphomas (PTLs) [1,8]. The patients may have retroperitoneal lymph node involvement at the time of presentation and an increased propensity to involve extranodal sites such as contralateral testis, central nervous system, lung, skin, and Waldeyer ring [2,3,9].

In general, testicular lymphomas are classified into pediatric and adult lymphomas. Pediatric lymphomas have a better prognosis and are mostly follicular lymphomas; on the contrary, adult lymphomas have a poorer prognosis and are 90% DLBCL [4]. These tumors can be graded as low, intermediate, and high grade [4]. Primary testicular DLBCL may present along with hydrocele in 40% of patients [5]. DLBCL has been reported as the most common bilateral tumor of the testis. Our patient had a unilateral testicular lesion, but the involvement of the contralateral testis may occur a few years after orchidectomy and can be a histologically different type [5].

Grossly, the tumors appear large, lobulated, fleshy to firm, homogenous, tan, or pale yellow with hemorrhagic and necrotic areas [5]. Microscopy reveals the tumor cells infiltrating as discohesive sheets in the interstitial spaces and sparing the seminiferous tubules. However, the tubules are filled up with tumor cells showing architectural effacement. The neoplastic lymphoid cells are medium sized to large with scanty basophilic to amphophilic cytoplasm, round to oval, and vesicular nuclei with fine chromatin. Few large lymphoid cells may have distinct nucleoli mimicking undifferentiated carcinoma [5].

IHC should be done to rule out non-seminomatous germ cell tumors of the testis and to classify this heterogeneous group of lymphoma. DLBCL is further subclassified into two groups – germinal center B-cell-like and non-germinal center B-cell-like. CD10+/Bcl6 positive and MUM1 negative are germinal center B-cell-like DLBCL and they are reported to have better survival rates. CD10/Bcl 6/MUM1 negative is non-germinal center B-cell-like with a worse prognosis and survival [10]. CD20 also shows diffuse strong positivity [11].

The patients are usually started on post-operative chemotherapy with a CHOP regime (cyclophosphamide, hydroxydaunorubicin, Oncovin, and prednisone) [3,4,9,11]. Since the tumor has been found to have very high rates of CNS relapse, prophylactic measures such as intrathecal methotrexate, cranial irradiation, and chemotherapy can be tried to reduce relapse [1,4,9]. Systemic dissemination also has been reported in many cases; hence, all the major lymphoid organs of the body particularly pelvic and mediastinal lymph nodes should be evaluated in detail. Aggressive management of these patients with post-operative chemotherapy, prophylactic CNS, and contralateral testicular irradiation is must. Even with vigorous treatment, the prognosis of the high-grade disease is <1 year [11].

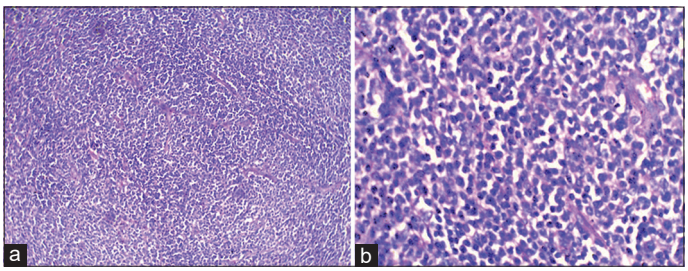

Figure 1: Photomicrograph shows (a) tumor cells in dyscohesive sheets with architectural effacement of tubules (×10) and (b) medium- to large-sized neoplastic lymphoid cells with round to oval vesicular nuclei, scanty basophilic cytoplasm (×40)

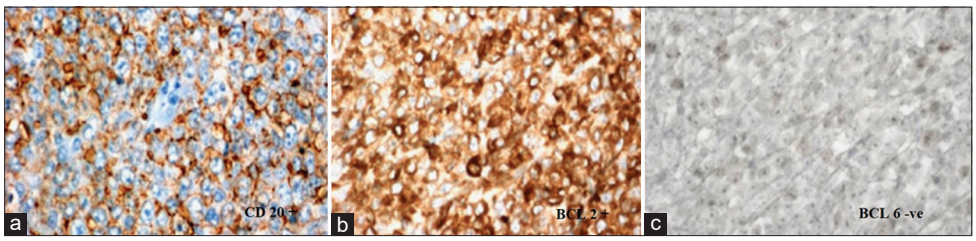

Figure 2: Immunohistochemistry photomicrograph showing (a) CD20-positive tumor cells; (b) BCL 2-positive tumor cells; and (c) BCL 6-negative cells

CONCLUSION

Primary malignant lymphoma is a rare testicular tumor and it contributes to <5% of all testicular neoplasms. The median age at diagnosis of DLBCL was 66 years (range 19–91 years) and 90% of lymphomas are of B-cell lineage. Testicular DLBCL has a high risk of extranodal relapse, even in cases with localized disease at diagnosis. Overall survival is 15–30% at 2 years. Anthracycline-based chemotherapy, CNS prophylaxis, and contralateral testicular irradiation seem to improve the outcome.

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