



Primary Testicular Lymphoma: A Case Report

Primer Testiküler Lenfoma: Olgu Sunumu

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ABSTRACT

Primer testis lenfoması nadir görülen bir tümör olup, bütün testis tümörlerin %1-9'unu oluşturmaktadır. En sık rastlanan histolojik tipi diffüz büyük B hücreli lenfomadır. Altmış yaşın üstünde en sık görülen testis malignitesidir. Prognozu diğer testis tümörlerine göre daha kötüdür. Prognozu belirleyen en önemli parametre evre olup orşiektomi sonrası adjuvant radyoterapi ve/veya kemoterapi uygulanmaktadır. Biz 50 yaşındaki bir olguyu literatür eşliğinde sunduk.

Anahtar Kelimeler

Testis, lenfoma, orşiektomi

ÖZET

Primary testicular lymphoma is a rarely seen testicular tumor accounting for 1-9% of all testicular tumors. The dominant histological subtype is diffuse large B-cell lymphoma. It is the most common testicular malignancy in men aged over 60 years. Its prognosis is worse than other testicular tumors. The most important parameter determining the prognosis is the stage. Adjuvant radiotherapy and/or chemotherapy are carried out after orchiectomy. In this study, we present the case of testicular lymphoma in a 50-year-old male patient and a review of the literature.

Key Words

Testis, lymphoma, orchiectomy

Introduction

Primary testicular lymphoma was first reported by Curling in 1866 (1). It is clinically aggressive and rarely seen, and though it accounts for 1-9% of all testicular malignancies and 1-2% of all non-Hodgkin's lymphomas, it is the most commonly observed testicular malignancy in men aged over 60 years (2). There is no standard treatment modality for primary testicular lymphomas due to their rare occurrence, however, today, chemotherapy and radiotherapy are performed after orchiectomy. When considering all stages, 5-year survival rate is 12% for these tumors showing frequent relapses (3).

Case Presentation

A 50-year-old patient applied to our clinic with the complaint of swelling in his right testis ongoing for fifteen days. On physical examination, a mass measuring 2 cm in diameter was palpable in his right testis, and the contralateral testis and other structures were found to be normal. A hypoechoic well-circumscribed mass lesion

showing internal vascularization within the testicular parenchyma was detected by color Doppler ultrasonography. Magnetic resonance imaging (MRI) demonstrated a large number of multiple lesions with the largest one measuring 1.5 cm in diameter within the testis with the presence of contrast and restricted diffusion (Figure 1). After carrying out necessary preoperative preparations for the patient in whom AFP, beta-HCG and LDH levels were normal, right radical inguinal orchiectomy was performed under general anesthesia. Histopathological examination revealed diffuse large B-cell lymphoma (Figure 2) and reported that the tumor was limited to the testicular parenchyma and lymphovascular invasion was not observed. Our patient is followed up by the medical oncology and radiation oncology departments with the purpose of adjuvant chemotherapy and radiotherapy.

Discussion

Histologically, 80-90% of the rarely seen primary testicular lymphomas are diffuse large B-cell lymphomas (4). Percent sixty of patients are in

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stage I while 30% are in stage II, and bilateral testicular involvement is seen approximately in 35% of cases (5). The most frequent symptom is painless testicular swelling fever, weight loss, night sweating, and fatigue; besides, loss of appetite can also be seen (6). Granulomatous orchitis, pseudolymphoma, plasmacytoma, and rhabdomyosarcoma are other conditions mimicking testicular lymphoma. Intratubular germ-cell neoplasia, the precursor lesion of testis tumors, is not present in primary testicular lymphoma (7).

Its etiology includes prior trauma, chronic orchitis, undescended testicles, and filariasis of the spermatic cord (8). Advanced stage tumors tend to spread to extra-nodal areas such as central nervous system, skin, Waldeyer's ring and the lungs (9). Classification of primary testicular lymphoma that was modified by the Nordic Lymphoma Group is as follows: stage I: unilateral testis involvement with or without epididymis or cord involvement; stage II: abdominal and pelvic lymph node involvement; and stage II-IV: distant metastasis (10).

The most important factors identifying prognosis are the clinical stage and histological grade (11). It has been reported that a primary tumor larger than 9 cm, epididymis, presence of spermatic cord and bilateral testis involvement, vascular invasion, advanced age, high LDH levels, presence of B symptoms, high International Prognostic Index (IPI) score, and left testis involvement are factors associated with poor prognosis (12). Nevertheless, young age, localized tumor, presence of sclerosis, small size of the tumor, low histologic grade and no epididymis or spermatic cord involvement are indicators of good prognosis (13). It is more frequently seen, and the prognosis is generally poor in HIV-positive men and in young ages (37 in average) (14).

Lymphomas are observed in the form of hypoechoic lesions on scrotal ultrasonography different than homogeneous hyperechoic normal testicular tissue. There is no standard treatment modality for primary testicular lymphoma as it is a rare incidence. In stage I and II cases, R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone) and prophylactic scrotal radiotherapy along with systemic chemotherapy are performed after orchiectomy (15). If adjuvant therapy is not administered after orchiectomy in stage I primary testicular lymphoma, distant relapse occurs in the majority of patients. In stage II cases, radiotherapy applied on localized lymph nodes can provide the prevention of nodal relapse. In stage III-IV diseases, systemic chemotherapy, scrotal radiotherapy, and intrathecal chemotherapy are performed. Prophylactic scrotal radiotherapy reduces the risk of relapse in the contralateral testis. It has been stated that the risk of relapse in the central nervous system, which is the most important complication that can be seen approximately within ten years after diagnosis, could be lowered by performing intrathecal chemotherapy (16). In our patient, a systemic chemotherapy protocol comprising the combination of rituximab, cyclophosphamide, doxorubicin, and vincristine and an intrathecal methotrexate therapy were implemented with the purpose of decreasing the risk of relapse in the central nervous system. Additionally, radiotherapy was conducted on the contralateral testis and on the paraaortic and right iliac lymph nodes, and no relapse was observed at the end of a 6-month follow-up.

In conclusion, primary testicular lymphoma is a disease with a poor prognosis. The rare incidence of the disease, its development and tumor behavior being different from the germ-cell cancers of the

testis have made it difficult to determine the treatment procedures to be applied after orchiectomy. Therefore, primary malignant lymphoma of the testis should be considered as the manifestation of systemic disease in the testis. The contralateral testis and relapse in the central nervous system should always be taken into consideration. Lymphoma

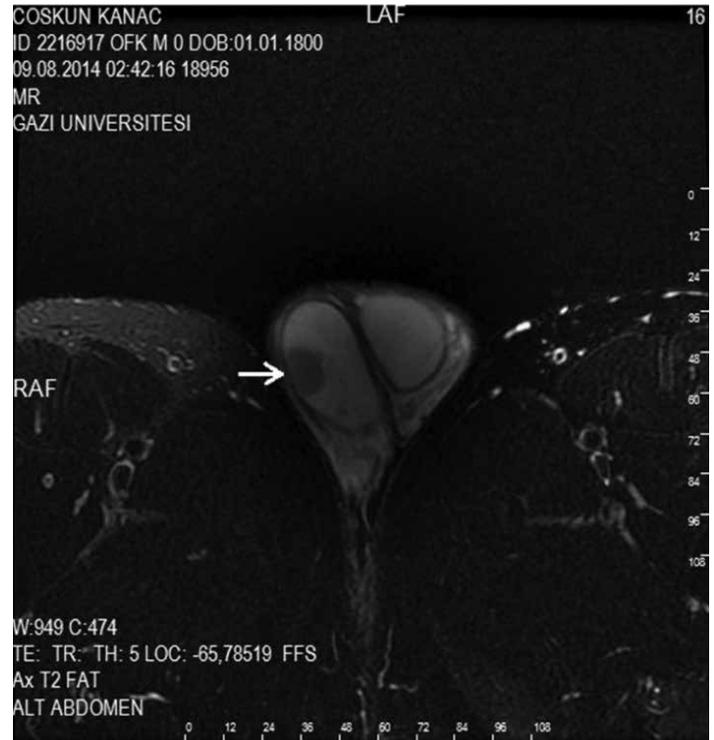


Figure 1. MR images of the primary intratesticular lymphoma (white arrows)

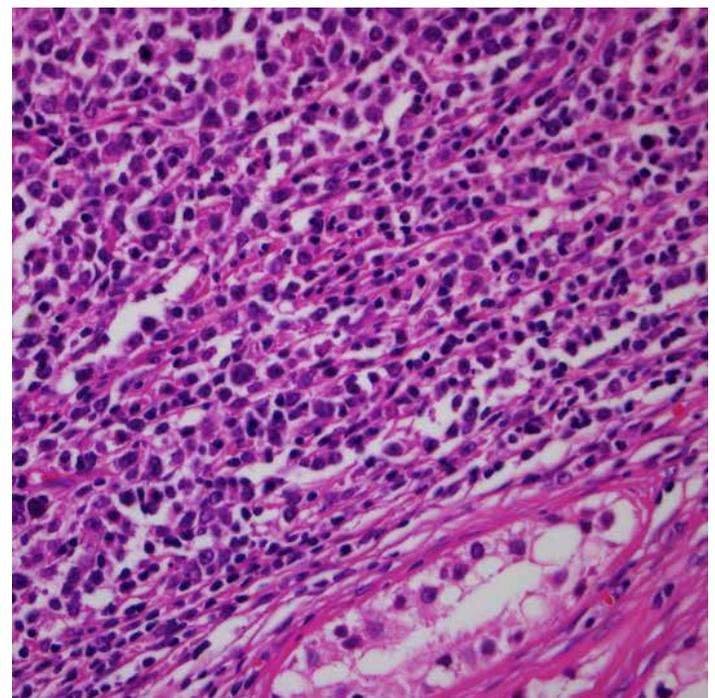


Figure 2. Atypical round cell tumor cells in the testis tissue, having lymphoid cell traits (HE x200)

should be kept in mind for patients who present with a mass in the testis, and the urologist, pathologist, and oncologist should take joint action.

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