

Synchronous Testicular and Orbital Involvement in a Child with Burkitt's Lymphoma

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Dear editor;

Burkitt's lymphoma (BL) is a mature high-grade aggressive B-cell non-Hodgkin's undifferentiated lymphocytic lymphoma. It is the fastest growing tumor with a doubling time of less than 24 hours.¹ It is common among children in Central African countries but it has also been reported in other geographic regions. Testicular involvement in non-Hodgkin's lymphoma (NHL) can be seen at initial diagnosis or during relapses. Orbital involvement is very common in endemic BL, but in contrast, it is very rare in sporadic form.² In this study we aimed to report on an immunocompetent child diagnosed with BL through synchronous testicular and orbital involvement.

A 13-year-old boy was admitted to our clinic with complaints of weakness for three days and swelling of his left eye and scrotum for seven days. The patient reported no recent illness, fever, weight loss or trauma. Physical examination revealed scrotal mass bilaterally, swelling of the left eye (Figure 1) and hepatosplenomegaly. On external ocular examination, the left eye was 9 mm proptotic, extraocular motility was severely restricted in all fields of gaze, and the upper and lower lids were severely edematous. His laboratory values were Hb: 7.9 g/dl, Hct: 22.9%, WBC: 25.9 x 10⁹/L with 76% polymorphs, 20% lymphocytes, 2% monocytes, 2% eosinophils, platelets: 137 x 10⁹/L, BUN: 35.3 mg/dl, creatinine: 0.68 mg/dl and lactate dehydrogenase: 510 U/L. Se-

rum electrolytes, serum immunoglobulins (including also anti EBV [Epstein-Barr Virus] antibodies), EBV Polymerase chain reaction and urinalysis results were normal. Scrotal ultrasonography (USG) revealed increase in both testes' size (the left testis was 18x16x36 mm and the right testis was 16x14x28 mm), bilateral heterogeneous testicular parenchyma and hypoechoic markedly hypervascular areas. Orbital magnetic resonance imaging (MRI) revealed plane mild diffuse thickening in the left periorbital area and a heterogeneous slight increase in this localization. Abdominal MRI showed hepatosplenomegaly and a nodular lesion showing contrast enhancement in the right kidney inferoposterior 28x33 mm in size.

Bone marrow aspiration was performed in the patient and L3 blastic cells (< 5%) were revealed. Lumbar puncture examination showed no evidence of central nervous system involvement. A testicular biopsy was performed. Microscopic examination showed medium-sized atypical lymphocytes with round nuclei, prominent nucleoli, many mitotic and apoptotic figures. The presence of scattered tangible body macrophages gave a starry sky appearance. Immunohistochemistry performed on tumor cells were positive for CD19, CD20 and CD79a. Although cytogenetic studies and Fluorescent in situ hybridization analysis were not specific for BL, this was the final diagnosis based on histological figures and immunophenotyping.



Figure 1. Clinical appearance demonstrating orbital and scrotal swelling

Patient was assessed as stage 4 BL and a B-NHL-BFM fourth risk group chemotherapy protocol was started. After first-line chemotherapy, the patient had dramatic improvement in proptosis and both testes' size almost decreased to normal size. After three months, the patient's bone marrow aspiration, scrotal USG and orbital MRI results were reported as completely normal. The patient did not have any complaints during the one-year follow-up and the illness was considered to be in remission.

Testicular involvement in NHL is usually present with disseminated disease. Kellie et al.³ noted in their review of 131 children with NHL that only six cases were admitted with the testicular involvement and that three cases had testicular relapse in follow-up. Thus, they reported the incidence of testicular involvement in NHL as 7%. Sonographic findings are not always specific enough to differentiate between benign and malignant lesions. However, USG is an excellent non-invasive method to evaluate scrotal masses.

While orbital involvement is very common in endemic BL, it is very rare in sporadic form. Baker et al.⁴ reported a review of the literature on children and adult patients with sporadic BL that was published on PubMed between 1959-2009. They noted three cases that were diagnosed and reported in a total of 16 patients experiencing orbital involvement with their ages ranging from 10 months to 84 years.

Lamm et al.⁵ reported on a 9 month-old boy who presented with left eye and testicular swelling and

redness of eyes; he had testicular biopsy results in accordance with BL. In the literature, our case is the second in which the illness is known to occur with both testes and orbital involvement.

In recent years, 75% of patients have been cured due to the improvements made in supportive care and multiagent chemotherapy, which is determined by histological subgroups and stages. The prognosis for patients whose illnesses are in advanced stages vary according to histological subgroups and survival has improved compared to previous years (event-free survival: 60-90%). Early diagnosis, staging and treatment play particularly important roles in ensuring remission in these patients' illnesses.⁶

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