Non-African Burkitt’s Lymphoma Manifesting at the Jaw and as a Right Orbital Mass in a Child

Sevin Söker Çakmak, MD
Muras Söker, MD
Orhan Ayıldız, MD
Celal Devecioğlu, MD
Sevda Ipek Söker, MD

INTRODUCTION

The non-Hodgkin’s lymphomas are a diverse collection of lymphoid malignancies that differ regarding pathology, cell of origin, natural history, and response to treatment. Burkitt’s lymphoma is a small, noncleaved, B-cell-derived lymphoma that is endemic in some parts of Africa. Non-African Burkitt’s lymphoma occurs sporadically in the United States and Europe.1 Burkitt’s lymphoma is more common in males and tends to present during the first two decades of life. Orbital lymphomas tend to be localized, and 10% to 15% are bilateral. The most common orbital lymphoma is Burkitt’s lymphoma.2 The abdomen is the primary site (more than 90% of cases) of non-African Burkitt’s lymphoma, with orbital involvement being extremely rare.

We report a case of histopathologically confirmed non-African Burkitt’s lymphoma with a rare presentation in an immunocompetent patient.

CASE REPORT

A 12-year-old boy with a primary manifestation of non-African Burkitt’s lymphoma in the right orbit presented to our department (Fig. 1). He had a 10-mm right proptosis and a regular, nontender mass involving the right upper eyelid. His best-corrected visual acuity was 1/10. An ocular examination disclosed severe nonaxial proptosis with inferotemporal displacement of the globe, chemosis, and restricted motility in the right eye. A firm mass was palpable superior to the globe. Pupillary reactions, the intraocular pressure, and the fundus were normal. No other abnormalities were found. The left eye was normal. Computed tomography scans showed a soft tissue mass in the right orbit with evidence of adjacent bone involvement (Fig. 2). An orbital incisional biopsy was performed. A biopsy specimen revealed that the mass contained small, noncleaved cells with coarsely clumped chromatin and nucleoli. The histopathologic findings were characteristic of non-African Burkitt’s lymphoma (Fig. 3). The tumor cells showed B-cell markers (CD20, CD45RB, and CD79).

A systemic examination revealed a 2-cm hepatomegaly. He had no other pathologic systemic signs. Abdominal ultrasonography and computed tomography revealed enlarged abdominal retroperi-
toneal lymph nodes and hepatomegaly. The findings on a computed tomography scan of the chest were normal. The results of a complete blood cell count were normal. The erythrocyte sedimentation rate was 15 mm/h. The results of bone marrow aspiration and biopsy were normal. Blood studies showed an increased blood lactate dehydrogenase level of 541 U/L (normal, 100 to 190 U/L) and an increased uric acid level of 11 mg/dL (normal, 3.5 to 6.4 mg/dL). The calcium level was normal. The results of tests for Epstein–Barr virus and human immunodeficiency virus were negative. The results of a lumbar puncture were normal.

The patient was diagnosed as having stage III disease according to Murphy staging criteria. He was treated with the NHL–BFM 90 (non-Hodgkin's lymphoma–Berlin–Frankfurt–Münster-90) regimen. After completion of initial chemotherapy (200 mg/m²/d of cyclophosphamide and 30 mg/m²/d of prednisolone for 5 days), the masses rapidly regressed (Fig. 4). Next, six cycles of chemotherapy (which included cyclophosphamide, vincristine, prednisolone, methotrexate, cytarabine, ifosfamide, doxorubicin, and etoposide) were completed. Six months after completion of chemotherapy, the patient remained in total remission.

**DISCUSSION**

Burkitt’s lymphoma is a monoclonal proliferation of B lymphocytes with a typically histologic pattern often called “starry sky.” All Burkitt and Burkitt-type lymphomas have surface markers of mature B cells. This lymphoma is one of the fastest growing malignancies. The involvement of the jaw or the orbit is uncommon in non-African Burkitt’s lymphoma, and no such cases were seen in one large study of pediatric orbital tumors.

The Epstein–Barr virus has been implicated in the African type, whereas a relationship between the virus and the non-African type is controversial. Non-African Burkitt’s lymphoma is common in immunodeficient patients.

Burkitt’s lymphoma is generally extranodal, with a predilection for the ileocecal region and mesentery in the non-African type and for the jaw in the African type. Although Burkitt’s lymphoma is clinically aggressive, it can be cured with multiagent chemotherapy. Burkitt’s lymphoma cells express pan-B-cell antigens, surface immunoglobulin of the
IgM type, and also CD10 antigen. The most frequent translocation—t (8;14) (q24;q32)—occurs in 80% of cases and juxtaposes the c-myc gene with one of the immunoglobulin loci on chromosome 14.

Patients with Burkitt’s lymphoma present with highly proliferative tumors, and frequently have peripheral and bone marrow infiltration. The involvement of the central nervous system is rare at diagnosis. Hyperuricemia and renal dysfunction may be found at diagnosis. The serum lactate dehydrogenase level is often elevated. Tumor lysis is common and requires special precautions. To prevent tumor lysis, some groups prefer to give an initial low dose “cytoreductive” treatment followed by more intensive chemotherapy.5

The prognosis for this disease is directly related to tumor burden. Involvement of the central nervous system and bone marrow is indicative of a poor prognosis. Younger age and focal disease at diagnosis are associated with a good prognosis.6 Previously, surgical resection was recommended for patients with localized disease. Currently, the mainstay of management involves chemotherapy. The alkylating agent cyclophosphamide was found to provide a cure in a few cases but is no longer used. Current modes of therapy include the combination of cyclophosphamide, vincristine, methotrexate, and prednisolone. Most patients receive intrathecal methotrexate and craniospinal radiation for central nervous system prophylaxis. Maintenance therapy is not necessary, and treatment is usually completed within 3 to 6 months. In refractory or recurrent disease, high-dose chemotherapy followed by autologous transplantation has been successful. Our patient had a favorable response after 5 days of cyclophosphamide and prednisolone and achieved total remission after completion of all cycles of chemotherapy.

This case supports the fact that non-African Burkitt’s lymphoma can manifest as acute proptosis and respond favorably to cyclophosphamide and prednisolone.

REFERENCES