

American Burkitt's lymphoma: report of a case with involvement of the jaws

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Abstract

The case of a nine-year-old Kansas boy with an American Burkitt's lymphoma is presented. The similarities and differences between the African and American variations of the disease are discussed. Included are descriptions of the diagnosis, clinical course, treatment of the lymphoma, and several common and uncommon characteristics of this case.

Burkitt's lymphoma is the most rapidly growing human neoplasm, doubling in size every 24 hours. It also has a growth fraction proportion of actively proliferating cells approaching 100%. The tumor was seen originally in patients in Africa; other cases have been reported in the United States, United Kingdom, Malaya, and Brazil.^{1,2} The disease is defined as a "malignant lymphoma, Burkitt's type, undifferentiated, and is comprised of uniformly primitive lymphoreticular cells."² Cytology shows characteristic round or oval malignant cells, 15-24 microns in diameter, with basophilic, vacuolated cytoplasm, and a round nucleus. Histologically, at low power the tumor is a mass of uniform-sized basophilic cells, often with pale histiocytes interspersed, giving a "starry sky effect."³⁻⁶

Background

The cause of Burkitt's lymphoma originally was thought to be the result of a vectored virus because of epidemiological evidence demonstrating climatic dependence and epidemic behavior. The virus consistently implicated was the Epstein-Barr virus first noted in cultured Burkitt's cells. Antibodies to the virus, though present in the general population of the world, consistently reach high titers in Burkitt's lymphoma patients. Etiology has been related to climate, altitude, and humidity.^{2-4,7}

Comparative studies of Burkitt's lymphoma in endemic areas of Africa and the United States have shown that many of the clinical features are similar throughout the world.⁸ Burkitt's lymphoma is found most commonly in

childhood, with a mean age in African cases of 9.1 years and in American cases of 12.2 years.^{9,10} The disease shows a preference for males in a 1.5:1 ratio. The predominant anatomic distribution in African cases in young patients is in the jaw and in older patients the abdomen. In American cases, abdominal involvement predominates; jaw involvement is relatively uncommon.⁹ The abdominal involvement usually affects the kidneys, ovaries, retroperitoneal tissues, and mesentery. Rarely, the presenting manifestations of Burkitt's lymphoma are tumors of bone (usually the distal femur), tumors of the thyroid, breasts, or soft tissues.^{3,4,7}

Both African and American cases of Burkitt's lymphoma demonstrate a good response to early high doses of cyclophosphamide.⁹ However, in the African cases two patterns of relapse may be seen. There may be an early relapse after a short remission. If this occurs, the disease is resistant to further treatment and the prognosis is poor. A second pattern of relapse after a long remission also may be seen. In this situation, the response to treatment is comparable to previously untreated patients.⁹ In American cases of the disease, only the early relapse pattern has been noted.⁹ A similarity between African and American cases is the curability of the diseases in a significant percentage of cases. The disease stage appears to be a more important prognostic factor in African cases.⁸ The stages are: Stage I — tumor confined to one anatomical site; Stage II — tumor involvement in two or more sites excluding the abdomen; Stage III — intra-abdominal tumor with or without jaw involvement, but without involvement of other sites; and Stage IV — intra-abdominal tumor with or without jaw involvement, but with tumor deposits in other sites as well.

In regard to epidemiology, both American and African cases of Burkitt's lymphoma demonstrate time-space clustering and an absence of cases from high altitudes. In African cases, the Epstein-Barr antibody titer is markedly higher while in American cases, the titer may be higher but not to the same degree.⁹

Clinical Report

B.C. was a nine-year-old Caucasian male who had lived his entire life in Kansas when he presented at the Pediatric Dental Clinic at the University of Kansas Medical Center. Six weeks prior to his first visit to the Medical Center, B.C. was taken to his family dentist because it "hurt when he chewed." He was told that his "permanent teeth were more mature than his age warranted" and that they were pushing out his deciduous teeth.

He had no increase in chewing pain until two days before his visit to the Dental Clinic. He was also beginning to develop bilateral knee pain. The family took B.C. to another dentist who referred him to a local physician; he was treated with erythromycin for five days. A panoramic radiograph was taken at that time. The patient then was referred to the University of Kansas Medical Center by the dentist and physician.

B.C. was an eight-pound, full-term, spontaneous delivery baby. There were no complications at birth and the gestation was normal. His development was within normal limits and he had no serious illnesses or injuries.

He was cooperative and in no apparent distress. Vital signs were as follows: pulse 92; respirations 32; blood pressure 120/88; and weight 30 kg. His head was normocephalic and his eyes were within normal limits. A 5 cm, mobile, painless, submandibular mass was noted on the right side of his neck. His cheeks were markedly swollen, distorting facial contours on the right side of his jaw. All teeth in the mandibular arch and the maxillary molars were mobile. The gingivae were hard with a marked increase in size (Figure 1). The throat was slightly

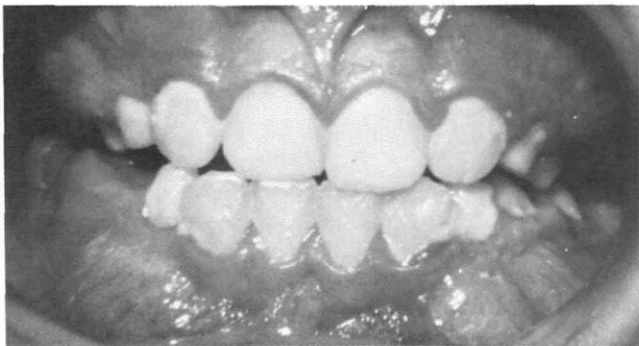


Figure 1. Gingival tissues at initial examination.

red and the tongue appeared normal. The chest was clear to auscultation. A slightly irregular heart rhythm with no murmur was noted. The extremities were within normal limits except for bilateral pain upon palpation of the knees and ankles.

The laboratory findings were as follows: WBC 11,600 with 59 segmented, 1 band, 34 lymphocytes, 3 monocytes, 2 eosinophils, 1 basophil; Hb 12.2; HCT 35.8; MCV 76.9; MCH 26.3; platelets 363,000; glucose 70; BUN 23; albumin 4.4; SGOT 55; cholesterol 157; CA

10.4; PO₄ 4.6; Na 142; K 5.2; Cl 105; CO₂ 17; LDH 2,916; PT 11.9; PTT 31.1. Radiographs of the upper GI and small bowel — esophagus, stomach, and entire intestine — were normal. Radiographs revealed multiple radiolucent lesions, primarily in the mandible (Figures 2 & 3).

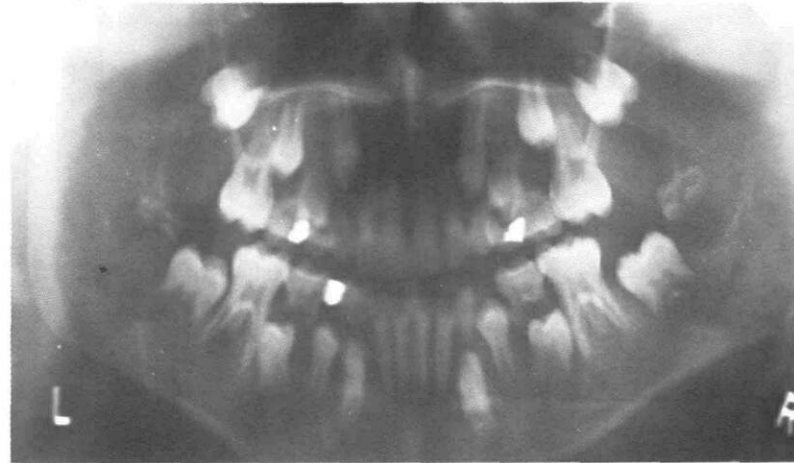


Figure 2. Panoramic radiograph at initial examination.



Figure 3. Bitewing radiographs at initial examination.

In order to determine the diagnosis, two definitive biopsies were performed. The dental staff performed a biopsy of the right mandibular gingival tissue. The ENT staff performed a biopsy of the right mandibular bone. Both pathology reports described the lesion as a diffuse lymphoma, Burkitt's type, Stage II. Also, during the initial hospital stay, bone scans were done in which the spine, left hip, mandible, and maxilla showed increased uptake of labeled phosphate, a radionuclide.

B.C. was started on a protocol which was being used by M.D. Anderson Hospital in Houston. This required an induction phase of vincristine (2 mg/m²) intravenous weekly for four weeks; prednisone (60 mg/m²) daily in four divided doses for 28 days, followed by a one-week tapering off period; cytoxan (1,200 mg/kg) intravenous on the first day only; and methotrexate (50 mg/kg) intravenous infusion for three consecutive weekly doses, then advancing to a dose of 100 mg/kg for one week.

The following week, 150 mg/kg of methotrexate was given and this continued for the next seven weeks, when a dose of 200 mg/kg was begun. Intrathecal treatment comprised another part of this therapy. Cytosine

arabinoside (45 mg/m²) was given intrathecally on days one and two; methotrexate (15 mg/m²) was given intrathecally on day three. This occurred on the first week and on the third week. The prognosis was guarded.

The patient was maintained on chemotherapy in six-week cycles for approximately one year. He was apparently tumor-free after six to seven months. He has not received chemotherapy for nearly two years and is still apparently tumor-free. The prognosis now is listed as good.

Discussion

Although this case closely followed the reported findings of American Burkitt's lymphoma, some differences were apparent; the patient's age and the primary involvement of the maxilla and mandible more closely resemble the African disease description. The early diagnosis and vigorous treatment of the disease according to the protocol of M.D. Anderson Hospital were keys to the successful treatment of the disease.

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