

# A pediatric case with disseminated Burkitt lymphoma presenting with facial soft tissue swelling and dental pain

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**A pediatric case with disseminated Burkitt lymphoma presenting with facial soft tissue swelling and dental pain**

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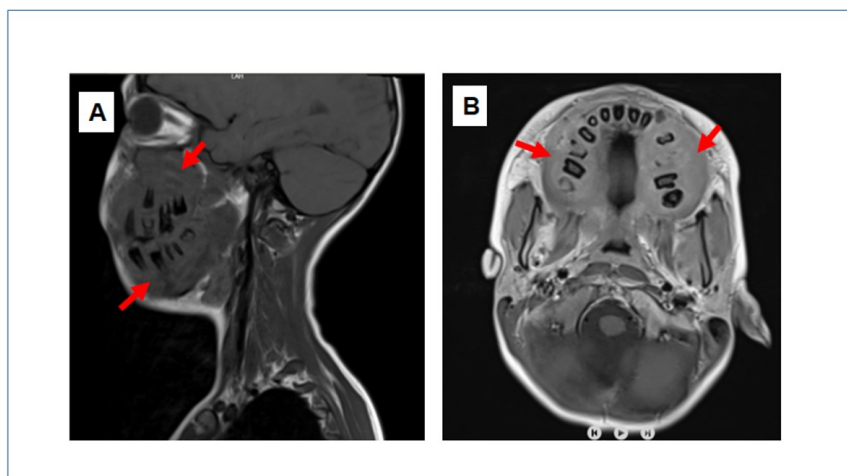
**Abstract:** Burkitt and Burkitt-like lymphomas constitute 40% of childhood lymphomas and are malignancies that require urgent diagnosis and treatment. Here, we present a case of a 10-year-old male patient who was diagnosed with Burkitt lymphoma and applied to the dentist with complaints of pain in the left upper molars and facial swelling for 2 weeks. Magnetic resonance imaging showed significant soft tissue swelling around the mandible and maxillary tooth roots in the lower and upper jaws. The patient was diagnosed with Burkitt lymphoma in the histopathological examination of the gingival biopsy. Diffuse bone marrow involvement and hyperuricemia due to tumor lysis syndrome were detected in this pediatric patient. The patient with stage IV Burkitt lymphoma has been successfully treated with intensive chemotherapy protocols and has been in remission for 5 years. Burkitt lymphoma should be considered in the differential diagnosis of soft tissue swellings in the jaw and orofacial region presenting with dental pain in childhood.

**Key Words:** Burkitt lymphoma, childhood cancer, dental pain

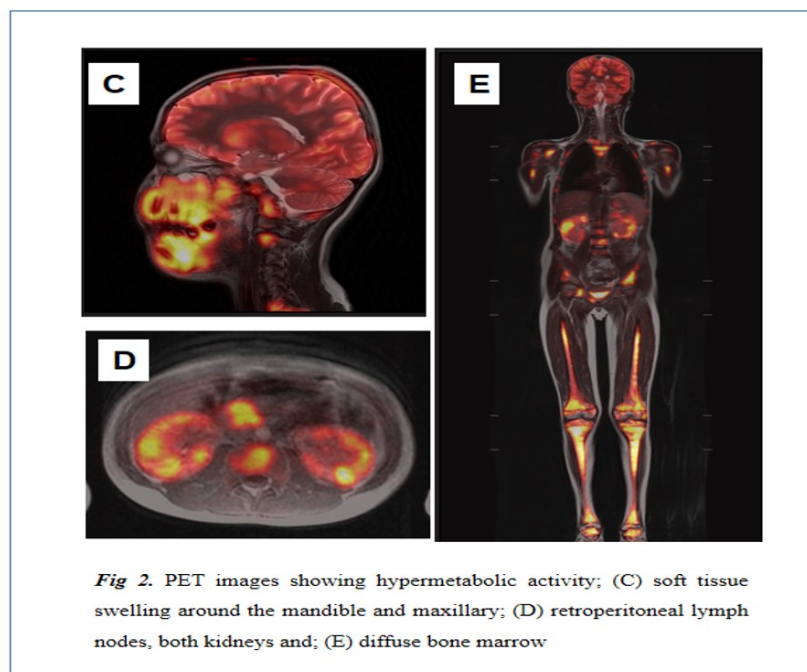
**Introduction:** Burkitt lymphoma, a subtype of non-Hodgkin lymphoma, is a malignancy that needs urgent diagnosis and treatment due to its high grade and growth rate in childhood. It is classified as endemic, sporadic, and immunodeficiency-related.<sup>1,2</sup> Disseminated disease, high tumor burden, and renal involvement can cause tumor lysis syndrome and can even result in death in patients with Burkitt lymphoma.<sup>3,4,5</sup> A boy child patient who applied to the dentist with facial swelling following toothache is presented here.

**Case Report:** A 10-year-old male patient applied to the dentist with complaints of pain in the left maxillary molars teeth and swelling on his face for 2 weeks. Tooth extraction was not performed and oral antibiotic treatment was started considering infection. Due to the swelling progressing toward the lower eyelid, a biopsy was taken from the gingival swelling, and the patient was quickly referred to our center. On physical examination, there was diffuse facial edema and swelling, and painless lymphadenopathies of 2x2cm were palpated in the bilateral upper cervical region. The child's past medical history was non-specific. The patient did not have any associated immunodeficiency and infectious agents (Epstein-Barr viruses and HIV) were negative. Hemogram and peripheral smear were normal. Although the biochemical values of potassium, phosphorus, urea, and creatine were within normal values, uric acid (8.0 mg/dl, normal: 3.5-6.5) and lactate dehydrogenase (LDH) (1710 mg/dl, normal: 150-350) levels were elevated in the patient. Magnetic resonance imaging (MRI) showed significant soft tissue swelling around the mandible and maxillary tooth roots in the lower and upper jaws (Fig.1A, B).

The patient was diagnosed with Burkitt lymphoma in the histopathological examination of the gingival biopsy. Histologically, there was a ‘starry sky’ appearance with cell morphology and immunohistochemical staining was diffusely positive for CD20, bcl-6, and CD10, and ki-67 staining was positive in more than 100% of the monomorphic neoplastic lymphoid cells in biopsy tissue. Imaging method performed for staging, whole-body positron emission tomography (PET)/MRI revealed hypermetabolic activity in soft tissue swelling around the mandible and maxillary, retroperitoneal lymph nodes, both kidneys, and diffuse bone marrow involvement (Fig.2 C, D, and E).



**Fig 1 A, B.** Soft tissue swelling around the mandible and maxillary tooth roots on MRI



**Fig 2.** PET images showing hypermetabolic activity; (C) soft tissue swelling around the mandible and maxillary; (D) retroperitoneal lymph nodes, both kidneys and; (E) diffuse bone marrow

Burkitt lymphoma involvement was detected in the bone marrow aspiration and biopsy. Finally, our pa-

tient was diagnosed with stage IV Burkitt lymphoma. The child was treated with the B-cell non-Hodgkin lymphoma treatment protocol of intensive systemic chemotherapy. Successful supportive treatment consisting of hydration, xanthine oxidase inhibitor (allopurinol), and urine alkalization with sodium bicarbonate were quickly applied for tumor lysis syndrome in our Hematology-Oncology Department. After 6 cycles of intensive chemotherapy treatment, the child patient has been followed in remission for 5 years.

## Discussion:

Burkitt and Burkitt-like lymphomas account for approximately 40% of childhood non-Hodgkin lymphomas.<sup>1</sup> Since Burkitt lymphomas are very rapidly growing tumors, the diagnosis should be made with the fastest and least invasive methods. A detailed history and complete systemic examination should be performed on each patient.<sup>5,6</sup> Soft tissue and bone infections, benign odontogenic cysts, and malignant tumors (Langerhans cell histiocytosis, Ewing sarcoma, non-Hodgkin lymphomas) should be considered in the differential diagnosis in childhood and adults.<sup>7,8</sup>

Burkitt lymphoma may show signs such as tooth displacement, dental loss, facial nerve palsy, and orbital swelling.<sup>9,10</sup> Patients who apply to the dentist with oral and dental problems and suspected Burkitt lymphoma should be urgently referred to pediatric hematology-oncology centers for diagnosis and treatment.<sup>11</sup> Complete blood count, peripheral smear, serum electrolytes (uric acid, phosphorus, urea, creatine, potassium), and LDH levels should be measured. Tumor lysis syndrome may develop in tumors such as Burkitt lymphoma with a high proliferation rate of tumor cells, elevated tumor burden (increased LDH, advanced stage, disseminated disease), and kidney involvement in cancer patients.<sup>10,12</sup> Our patient had a high risk of tumor lysis syndrome with elevated LDH levels, disseminated stage IV disease with bone marrow involvement, and renal involvement. There is no central nervous system involvement in this patient. Tumor lysis syndrome is a life-threatening condition characterized by organ dysfunction and metabolic problems such as hyperuricemia, hyperphosphatemia, hyperkalemia, and hypocalcemia.<sup>2,5</sup> Precautions should be taken in patients at high risk of developing tumor lysis syndrome or if it has developed, it should be treated urgently. Hydration, urine alkalization and drugs to lower uric acid levels should be given. The high levels of potassium released by the breakdown of tumor cells are life-threatening and should be treated urgently. Hemodialysis may be required in renal involvement without urine output, and metabolic disorders unresponsive to drugs. Our patient's laboratory findings improved with medical treatment (hydration, allopurinol), adequate urine output was achieved, and he did not need hemodialysis. Burkitt's lymphoma intensive chemotherapy protocol was applied to our patient and treatment-related side effects (mucositis, myelosuppression, febrile neutropenia,...) were successfully treated. Our patient with stage IV diffuse bone marrow involvement was successfully treated and remission was achieved. The prognosis is good even in advanced disease with intensive treatment regimens in Burkitt lymphoma in childhood.<sup>5,12</sup>

## Conclusion:

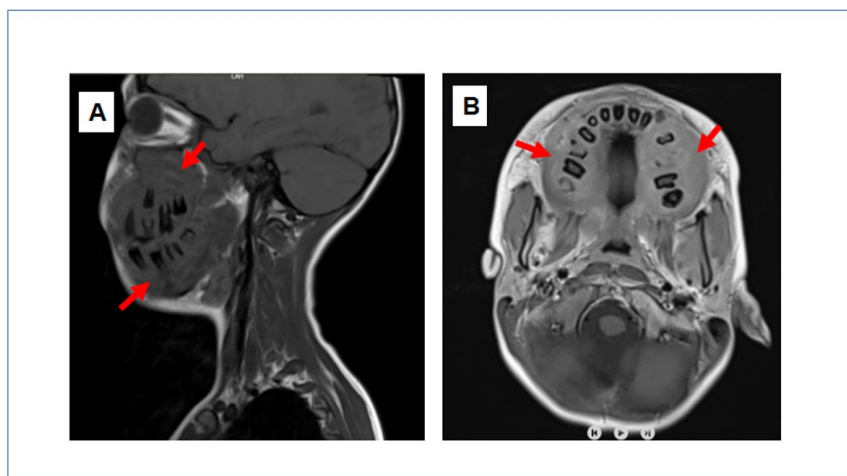
Although Burkitt lymphomas are rapidly proliferating, aggressive, high-grade lymphomas, the first sign may be extranodal involvement such as toothache, and facial soft tissue swelling. Burkitt lymphoma should be considered for tooth, jaw, and orofacial soft tissue swellings in the differential diagnosis, and early diagnosis and urgent treatment are life-saving in childhood and adult cancers.

**Conflict of interest:** None.

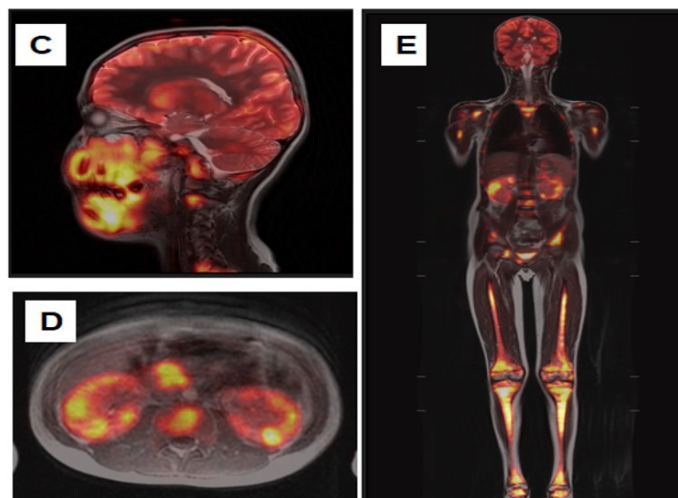
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