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ORIGINAL ARTICLE

Burkitt's Lymphoma In 3 Years-Old Boy With Craniofacial Presentation

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Background

Burkitt's lymphoma (BL) is an aggressive non-Hodgkin B-cell lymphoma with rapid growth. The majority of endemic BL occurs in children aged 5-12 years old with mandibular involvement.

Case

We presented a rare case of BL in 3 years-old boy with craniofacial involvement in Indonesia. A boy came to the emergency department with a massive mass in the upper-left facial area. The mass was 10 x 5 x 5 cm with eye and maxillary infiltration. Three months before, the patient came to the outpatient clinic with a slightly-visible and painless mass in the left cheek without any lymph node enlargement. The blood examination revealed no abnormalities, yet the LDH level was 1.260 U/L. The histopathological examination supported the diagnosis of Burkitt's lymphoma. No leukemic cells or metastases were found in bone marrow aspiration. The patient received Cyclophosphamide, Vincristine, Methotrexate, and oral prednisone. The patient responded well and the mass was completely resolved in 3 weeks with no complications.

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Figure 1 *(A) Initial presentation at the outpatient clinic; (B) Massive tumor on 5th day; (C) the tumor was significantly reduced on day 10th*

Burkitt's lymphoma (BL) is a rare aggressive high-grade B cell lymphoma with a rapid doubling time of 24–48 hours.[1] In Indonesia the incidence of BL is unclear due to limited tools and resources for diagnosing BL[2,3] Medical records from our hospital in Surabaya Indonesia revealed only 5 cases of BL from 2015 to 2019. The majority of BL occurrs in children aged 5-12 years old with jaw involvement.[4] We reported a case of 3 years old boy diagnosed with BL and craniofacial involvement.

CASE REPORT

A three years-old-boy came to the emergency department with a massive mass in the upper-left facial area. The mass was aggressively enlarged 3 months ago followed by severe pain 2 weeks ago. Fever hypersalivation, and sore throat were denied. The nutritional intake was decreased with significant weight loss. Three months before, the patient came to the outpatient clinic with a slightly-visible and painless enlargement in the left cheek without any lymph nodes involvement (Figure 1A). However the parents rejected hospital admission during the outpatient visit.

Based on physical examination, the mass was 10 x 5 x 5 cm, solid, and fixed without signs of inflammation. The mass had been infiltrated the eye and maxilla causing partial obstruction of the mouth. Minor gum bleeding was observed at the mass. Cervical submandibular, and submental lymph nodes were enlarged. No abnormalities were found in the chest, abdominal, and extremities. Neurological examination was normal.

Laboratory examination upon emergency admission showed no abnormalities (hemoglobin 10.9 mg/dL), yet the lactate dehydrogenase (LDH) level was 1.260 U/L The rapid HIV test was non-reactive. Peripheral blood smear revealed heterogenous normochromic-microcytic erythrocytes; impression of lymphocyte dominance with atypical lymphocytes; and increased platelet count. The patient was planned for fine-needle aspiration biopsy (FNAB) and bone marrow aspiration (BMA).

On the 5th day of admission, a massive gum bleeding (50 mL) from the mass occurred. The general condition was weak with poor oral intake. The hemoglobin and hematocrit were reduced to 9.5 mg/dL and 27.1%, respectively. The FNAB result demonstrated a malignant round cell tumor supporting BL (Figure 2). No malignancy cells were observed on BMA

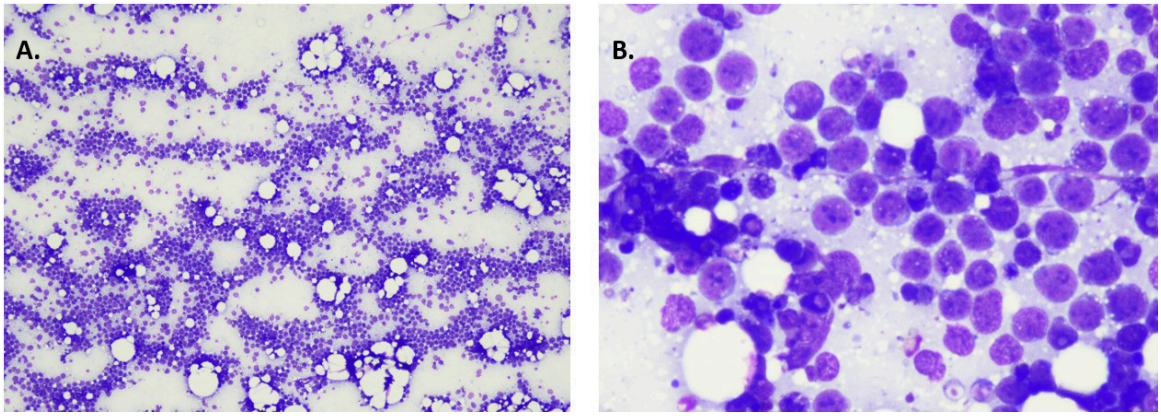

Figure 2 *FNAB finding (A) hypercellular smear with homogenous round anaplastic cells; (B) intermediate-sized anaplastic cells with round nuclei containing coarse chromatin, thin cytoplasm, and some lipid vacuoles.*

The patient was diagnosed with BL After bleeding control and transfusion, the patient received chemotherapy with Cyclophosphamide Vincristine Methotrexate and oral prednisone. The intake was partially supported by parenteral nutrition. The patient responded well, the mass size was reduced after the first regimen of chemotherapy. The nutrition could be administered orally. The LDH and uric acid levels were 480 U/L and 1.1 mg/dL, respectively. No abnormalities were found on the liver and kidney function test. After 3 weeks, the mass was completely resolved without any complication (Figure 1C). The patient was stable and went to the hematology outpatient clinic every 2 weeks.

DISCUSSION

Burkitt's lymphoma is an aggressive high-grade B cell lymphoma with rapid mass progression.[1] Burkitt's Lymphoma is classified into three types, endemic, sporadic, and immunodeficiency-related. The endemic type is the most common, especially in children. The clinical symptoms include typical painless mass around the jaw with/without intra-oral extension such as dental problems. The BL often occurs in males aged 5-12 years old, with peak incidence near 10 years old.[4,5] Our case demonstrated that endemic BL could occur in children aged under 5 years old with craniofacial involvement.

The patient experienced significant weight loss and nutritional problems which are typical for endemic BL patients.[1] In this patient, the significant weight loss is most likely due to the progressive enlargement of the tumor causing partial obstruction of the mouth and intake limitation. In addition, the high energy requirement for cancer cell proliferation also caused energy deprivation and malnutrition among BL patients.

The diagnosis of this patient was based on the FNAB result which supported BL findings. Histologically Burkitt Lymphoma is characterized by a diffuse growth pattern without any nodularity. The microscopic features of typical BL are homogenous intermediate-sized cells with round nuclei containing coarse chromatin, and multiple small nucleoli admixed with tingible body macrophages creating a "starry-sky" pattern visible at low power. Mitotic and apoptotic activity is typically a prominent feature.[6] Early pathological finding leads to early diagnosis which remains a key factor in pediatric oncology as it allows for early detection and proper treatment.

From the laboratory finding, the hemoglobin was normal during the first admission to the emergency department (10.9 mg/dL), yet major gum bleeding occurs on the 5th day causing a significant reduction of the hemoglobin (9.5 mg/dL). However after bleeding control and transfusion, the hemoglobin was recovered to 15.4 mg/dL It is plausible that the bleeding source was from the BL since BL has a rapid doubling with angiogenesis activity.[7]

The first-line treatment for all types of BL is intensive-short courses of chemotherapy combinations such as cyclophosphamide, vincristine, prednisone, doxorubicin, alkylators, and etoposide. No surgical or radiotherapy intervention is needed in managing BL Given the high dose and intensive chemotherapy administered during BL treatment, drug toxicities and complications are common. In addition, tumor lysis syndrome is a potential complication due to the rapid suppression of tumor cells.[1,6] Fortunately the patient responded well to chemotherapy. The uric acid after chemotherapy was normal, indicating no tumor lysis syndrome occurred.

Despite massive tumor size, bleeding, and nutritional problem, the patient had a good prognosis since the patient completely responded to chemotherapy. The tumor was not visible, hemoglobin was recovered, and adequate oral intake. The survival rate for BL had been improved over the years since numerous advances in diagnosis and treatment. Patients aged 19 years old or less have the best prognosis with 87% 5-year survival, while older patients and those with advanced disease have the worst prognosis.[6]

CONCLUSIONS

Burkitt's lymphoma is a rare tumor in children aged under 5 years old. Early clinical and pathological examination is the key factor for diagnosing BL.

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