

CASE REPORT



Oral manifestations of acute leukemia in children: family supernatural beliefs and its contribution to the prognosis of the disease

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Abstract

Aim: This case report aims to elaborate on the oral manifestations of acute leukemia in children and the extent to which the family's supernatural beliefs contributed to the prognosis of the disease.

Background: Leukemia is a malignancy of hematopoietic cells, and about 56% of cases are manifest in the oral cavity. In Indonesia, cancer is among diseases often related to a supernatural phenomenon that may hinder the patient from getting medical treatment.

Case Description: An 8-year-old girl was referred to the Oral Medicine Department due to swollen and bleeding gums. There was generalized enlargement of the gingiva with a reddish-purple color covered 2/3 of the dental crown and easy to bleed. The patient was diagnosed with gingival enlargement and gum bleeding due to acute leukemia as the suspected underlying disease. The patient was scheduled for a bone marrow aspiration examination to confirm the blood disorder diagnosis, but the family refused it and believed that supernatural power was the cause of the cancer.

Conclusion: Gingival enlargement and spontaneous bleeding of the gum are often the first oral signs of acute leukemia. Considering the influence of supernatural beliefs in society regarding cancer is still existing, dentists are required to be more persistent in educating patients and their families to receive definitive therapy to increase patient life expectancy.

Clinical Significance: The oral manifestation of leukemia should be treated promptly to support the systemic therapeutic. Delay in diagnosis and treatment will lead to poor outcomes of the disease.

Keywords: Beliefs, children, Indonesia, leukemia, oral manifestation

Introduction

The Global Burden of Cancer 2020 (GLOBOCAN 2020) reports that 19.1 million new cancer cases occurred in 2020, with the incidence in children and adolescents aged 0–19 years is no less than 400,000.^[1] Survey shows that one in three cancers that occur in children is leukemia.^[2] In Indonesia, of the 396,914 incidences of cancer, 14,979 cases were leukemia, with a mortality rate of 11,530.^[3] The four most common types of leukemia are acute lymphocytic leukemia (ALL), chronic lymphocytic leukemia, acute myeloid leukemia (AML), and chronic myeloid leukemia, and of the four types, which mainly affect children are ALL and

AML with a prevalence ratio of 6:5.^[2] The etiology is unknown but it has several risk factors, such as genetics, gender (men are more susceptible than women), age (certain types of leukemia have different risks), genetic disorders, lifestyle (smoking, alcohol, and obesity), radiation, chemotherapy, chemicals, and electromagnetic waves.^[2]

When a person experiences cancer, the impact is not only on the physical but also on the emotional and financial condition of the sufferer, his family, community, or even the health system. Health behavior in low- and middle-income countries, including Indonesia is still full of myths, stigmas, and taboos related to cancer, which affect cancer awareness and ultimately contribute

to mortality rates.^[4] The impact of this phenomenon on cancer mortality needs to be studied further.

This case report aims to provide an overview of the oral manifestations of acute leukemia in children and the extent to which family supernatural beliefs contribute to the occurrence of the disease and the prognosis of cancer itself.

Case Report

An 8-year-old girl was referred to oral medicine department with the main complaint of swollen and bleeding gums, followed by weakness, pallor, joint pain, and bruises on the body, especially on the legs. Previously, she was examined by a dentist who noticed that her gingival enlargement was a possible manifestation of an underlying systemic disease.

The patient had anemic conjunctiva, lymphadenopathy, also a dry and desquamated lips [Figure 1a]. There was spontaneous gingival bleeding, generalized gingival enlargement with purple-reddish coloration with around 2/3 encroachment on anatomic crown [Figure 1b and c], and palatal pallor with extensive ecchymosis [Figure 1d]. The serological test showed a very high leukocyte count reaching 109.690/ μL , while the other parameters, on average, showed a decrease from normal values [Table 1].

The working diagnosis was gingival enlargement and bleeding caused by suspected ALL with AML as the differential diagnosis. The pediatrician provided hydration using dextrose monohydrate 75 cc/h, and transfusion of packed red cell optipress® and thrombocyte concentrates. The patient was instructed to maintain oral hygiene using gauze moistened with 0.9% NaCl, tranexamic acid mouthwash, 0.12% chlorhexidine digluconate, and ibuprofen syrup. After 4 days of treatment, some of the serological parameters slowly showed improvement [Table 1]. The patient is scheduled for a bone marrow aspiration to confirm the diagnosis of blood disorders. Unfortunately, the family refused it. We had informed the family regarding the

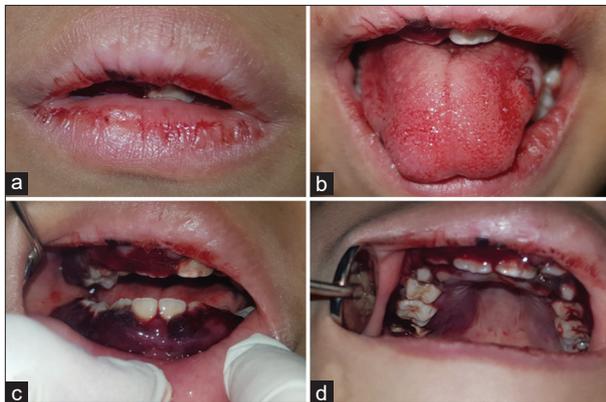


Figure 1: Extra and intraoral examination. Lips are dry and desquamated, and blood clots appear on the upper and lower lips (a), bleeding from gums on the dorsum of the tongue (b), gums are enlarged and bleeding (c), and the palatal mucosa was pale with a large area of ecchymosis (d).

oral conditions as the typical signs of leukemia, the urgency of diagnosis determination and immediate therapy, and also the prognosis of the disease. However, they were more convinced that the child was a victim of black magic, and choose to seek treatment from a medicine man. One month after hospitalization, the family told us that the patient had passed away.

Discussion

ALL is common in Hispanic and white races, aged 2–5 years, and is more frequent in boys, while AML is more common in 0–2 years old and adolescents, regardless of race and gender.^[2] Both types of leukemia have a poor prognosis, especially AML.^[5] Considering that leukemia has many subtypes, a bone marrow examination is required to confirm the type of leukemia and start an adequate therapy.

Signs and symptoms of leukemia are varied according to the type of neoplastic cell proliferation and their impact on normal cells: A decrease in erythroblasts causing anemia, pallor, fatigue, and arthralgia; decrease in granulocytes allows persistent infection and fever; while a decrease in platelet count shows petechiae, ecchymoses, and thrombocytes value of $2.5\text{--}6.0 \times 10^4 \text{ mm}^3$ causing spontaneous bleeding, which can be life-threatening.^[6] Platelets with leukemic cells can invade other organs resulting hepatosplenomegaly, lymphadenopathy, neurological disorders (trigeminal neuralgia, dysphagia, and trismus), and gingival enlargement.^[7] In line with this theory, the patient's serological results showed a decrease in erythrocyte, platelet, and granulocyte [Table 1].

Table 1: Serological examination

Parameter	Value		Reference value	Unit
	Initial examination	Last examination		
Hemoglobin	4.3 ^{LL}	8.8 ^L	11.5–15.5	g/dL
Hematocrit	16.1 ^{LL}	28.4 ^L	35–45	%
Erythrocytes	2.1 ^L	3.61 ^L	4–5.2	Jt/ μL
MCV	76.7 ^L	78.7	77–95	fL
MCH	20.5 ^L	24.4 ^L	25–33	Pg
MCHC	26.7 ^L	31	31–37	%
Leukocytes	109.69 ^{HHH}	40.76 ^{HH}	4.5–13	$10^3/\mu\text{L}$
Thrombocytes	9 ^{LL}	31 ^{LL}	150–450	$10^3/\mu\text{L}$
Total Neutrophils	2.19	0.82 ^L	2.10–8.89	$10^3/\mu\text{L}$
Total Eosinophils	0 ^L	0L	0.01–0.40	$10^3/\mu\text{L}$
Total Basophils	0 ^L	0L	0.01–0.09	$10^3/\mu\text{L}$
Total Lymphocytes	17.55 ^H	6.52 ^H	1.26–3.35	$10^3/\mu\text{L}$
Total Monocytes	12.07 ^H	1.63 ^H	0.29–0.95	$10^3/\mu\text{L}$

^{LL}: Very low, ^L: Low, ^H: High, ^{HH}: Very high, MCV: Mean corpuscular volume, MCH: Mean corpuscular hemoglobin, MCHC: Mean corpuscular hemoglobin concentration

Oral lesions could arise in around 65% of all types of leukemia, but primarily in the acute stage, especially in 90% AML,^[5] with 66.7% have abnormalities in the gingiva,^[7] and therefore can be an important diagnostic indicator. It may be the result of direct infiltration of leukemic cells or secondary to underlying thrombocytopenia, neutropenia, or impaired granulocyte function. Some of the oral manifestations reported in acute leukemia include mucosal pallor, petechiae, spontaneous bleeding, mucosal ulceration, gingival enlargement with or without necrosis, infection, hemorrhagic bullae on the tongue, and chapped lips.^[5,7] All these oral manifestations, except hemorrhagic bullae, were found in our patient [Figure 1].

Oral hygiene was maintained using gauze moistened with normal saline as an adjuvant strategy to reduce infection in the oral cavity. The sodium chloride crystalloid liquid (NaCl) 0.9% is an isotonic concentration of NaCl with a pH ranging from 4.5 to 7 which minimizing infection by increasing the function of macrophages (inducing the release of cytokines and chemokines) and pro-inflammatory T cells while reducing the potential risk of excessive regulation and anti-inflammatory action.^[8] Patient had also prescribed 0.12% chlorhexidine digluconate mouthwash. It has a bactericidal effect at high concentrations and bacteriostatic at low concentrations (below 4%).^[9] Ibuprofen syrup is also prescribed as an analgesic to treat acute pain. As a nonsteroidal anti-inflammatory drug, Ibuprofen has a better safety level than aspirin and higher efficacy than the acetaminophen group.^[10]

Tranexamic acid [4- (aminomethyl) cyclohexane carboxylic acid] mouthwash was prescribed to stop the gum bleeding. The Lysine-derived amino acid works as antifibrinolytic by binding to plasminogen, thereby preventing plasmin formation and the binding of plasminogen to fibrin, also stabilizing blood clots. It is effective for gingival bleeding in patients with leukemia, post-extraction, and mucosal bleeding.^[11] Controlling the spontaneous gingival bleeding will prevent the worsening of the systemic condition. Unfortunately, the patient was taken home before the medication was administered.

Examination of spinal cord aspiration is the key to diagnosis, definitive therapy, and prognosis of the disease.^[12] Compared to cytogenetic or immunohistochemical analyses, this technique has high reliability, and relatively affordable. Therefore, suitable for developing countries such as Indonesia. Sadly, the examination was not carried out, and the therapy had not given. Failure to achieve treatment for patients is one of the factors that can affect life expectancy.

The life expectancy of cancer patients is influenced by early detection, quality care, and care for cancer survivors.^[13] In developing countries, including Indonesia, the percentage of life expectancy for children and adolescents with cancer is 10–30%, in contrast to developed countries which can reach 80%. Life expectancy for 5 years or more after diagnosis is more significant in children aged 14 years or less, which is 90% in ALL and > 65% in AML.

Human belief in magic and its relation to disease and death has existed for centuries in almost all parts of the world. Research conducted in Indonesia shows that socio-cultural aspects of

society, such as belief in myths and magical power, greatly influence health behavior, both on prevention and treatment.^[14] A study conducted by Saeed *et al.* in 2019 showed that 55.5% of 402 cancer patients admitted that their cancer came from magic.^[15] This belief is generally driven by a fear of death, denial, and guilt.^[4] This is what happened to our patient's family, which claimed that their children were under influence of witchcraft, and considered alternative therapy is more promising than modern medicine. The failure of modern medicine to treat cancer, especially at an advanced stage makes people seek other options which they think is safer, more effective, and low-cost. However, the efficacy has not been empirically proven.

Delay in diagnosis, inability to get an accurate diagnosis, failure to access the right therapy, and neglecting treatment are aspects that impede the recovery and life expectancy of leukemia patients. It is undeniable up to date cancer is still attributed to myths and stigma. Therefore, it is necessary to educate the public to change health behavior, especially regarding acute leukemia in children.

Conclusion and Clinical Significance

The common and prominent oral manifestation of acute leukemia in children is gingival enlargement and gum bleeding, which are crucial signs for a dentist in the early detection of leukemia. In addition, supernatural beliefs regarding leukemia are a challenge for the dentist to be more persistent in educating patients and their families to get a definitive diagnosis and receive adequate therapy to increase patient life expectancy.

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