Management of aphthous-like ulcer induce by trauma in patient with thalassemia beta intermedia: a case report

Dahlia Riyanto,* Bagoes Soebadi, Priyo Hadi, Desiana Radithia

Abstract

Objective: Thalassemia is an inherited blood diseases cause which abnormalities systemically and intraorally. This case report to discuss management of Aphthous-Like Ulcer in 24 year old man with Thalasemia β Intermedia and need blood transfusion every month. Ulcer occurred 2 days before transfusion.

Methods: Based on history, clinical examination, hematologi and ferritin tests.

Results: The diagnosis was Aphthous Like ulcer induce by trauma. Patient was treated using anti-inflammatory mouthwash and recovery was achieved within 3 weeks follow-up.

Conclusion: Management of oral manifestation in Thalasemia patient need multidisciplinary approach to obtain successful therapy.

Keywords: Aphthous-like ulcer, Oral ulcer, Thalassemia β-intermedia


Introduction

The term “thalassemia” comes from the Greek word “Thalassa” which means sea and “Haema” which means blood. Thalassemia is one of the most common autosomal recessive blood disorders has been found in several countries with the main distribution in the areas known as the Thalassemia Belt, such as the Mediterranean, Middle East, Southern Asia, China Peninsula, Pacific Islands and Southeast Asia, including Indonesia with an incidence of 5-20%. Thalassemia is caused by a mutation in the gene responsible for the production of hemoglobin. Hemoglobin is a protein in red blood cells that carries oxygen, consisting of four protein chains, two globin chains α and two globin chains β arranged into heterotetramers. β-thalassemia occurs in two ways: transfer of recessive genes from parents to children and mutations in the Hb β gene on the autosomal chromosome No. 11. The severity of the disease depends on the number of gene mutations present in the patient. According to its severity, β-thalassemia is classified into three types: thalassemia major (TM), thalassemia intermedia (TI), and thalassemia minor. Mutations are characterized as β0 or β+. This β0 thalassemia is known as major beta thalassemia where no beta chain production is found and β+ thalassemia is known as intermediate beta thalassemia where beta chain production is decreases.

The globin chain imbalance causes hemolysis, which causes mild to severe anemia characterized by hypochromic and microcytic erythrocytes. Patients with anemia may have atrophic glossitis (AG) or generalized oral mucosal atrophy, recurrent aphthous ulceration (RAU) and tenderness or burning sensation in the oral mucous. RAU is clinically similar to RAS (Recurrent Aphthous Stomatitis) but does not begin in childhood or is associated with other features not related to classical RAS. Presentations like this are called Aphthous Like Ulcers (ALU).

Case Report

A 24-year-old male patient came to Airlangga University Dental Hospital on November 30th 2018, complaining of stomatitis because of being bitten when eating crackers 4 days ago and have not been treated. Patient admit that he often have stomatitis since +/-1 year ago, usually appearing once every month in different places when the patient is exhausted, stressed or because of a low Hb. The patient had a medical history of Intermedia β Thalassemia since the age of 1.5 years and underwent routine blood transfusion treatment once a month. The patient transfusion was 2 days ago with 4 bags of blood. Patient had a medical history of antrain drug allergy. Patient had been treated with folic acid and Feriprox once a day. A medical family history, the patient claimed his father had a history of anemia and hypertension. Results of a complete blood test before transfusion
Figure 1 Ulceration in the right labial mucosa (first visit), B. Erosion, solitary, in the right labial mucosa (7 days follow up), C. Erosion, solitary in the lower right labial mucous (14 days follow up), D. Macule, solitary, in the right labial mucous (21 days follow up)

Seven days later, the patient claimed that stomatitis in the right labial mucous had improved and painless. Patient explained that he used topical drugs according to the recommendations and used of the drug last December 7th, 2018 at 08.00. On intra-oral examination erosion was found, solitary at the right lower labial mucous, oval, 7x5mm in size, white, the area around redness, smooth surface, diffuse border, irregular edges, painless figure 1B. Working diagnosis was concluded as Aphthous-Like Ulcer. CIE including the use of drugs according to recommendations, adequate hydration, and maintaining oral hygiene.

After seven days, the patient claimed that stomatitis in the right labial mucous had improved and painless. Patient explained that he used topical drugs according to the recommendations and used of the drug last December 7th, 2018 at 08.00. On intra-oral examination erosion was found, solitary at the right lower labial mucous, oval, 7x2mm in size, white, the area around redness, smooth surface, diffuse border, irregular edges, painless figure 1B. Working diagnosis was concluded as Aphthous-Like Ulcer. CIE including the use of drugs according to recommendations, adequate hydration, and maintaining oral hygiene.

After another seven days, the patient claimed his condition has improved considerably. Stomatitis on the right labial mucous has healed and no pain. He use the last medicine this morning at 8:00. He explained that he used topical drugs according to the recommendations. The introral examination found macule, solitary, on the right labial mucous, oval, 4x1mm in size, white, the area around redness, smooth surface, diffuse borders, irregular edges, no pain figure 1D. On this visit the Aphthous like ulcer has healed and treatment was complete. CIE including continuous medication from internal medicine specialist routinely, diligently follow schedules for transfusions, maintain body condition, remain optimistic about medication, nutritious food and adequate rest and control if any more complaint in oral cavity.

Discussion

Thalassemia Intermedia (TI) is a term developed to describe patients with manifestations that are too mild for considered thalassemia major and too severe to be called minor thalassemia. The beta-thalassemias, including TI, arise from damaged gene function in partial suppression of beta-globin protein production.9

People with thalassemia produce more abnormal forms of globin chains and Hb compared to normal individuals. Imbalance of globin chains causes hemolysis, which causes mild to severe anemia. Ineffective erythropoiesis happens when alpha chain is very unstable and precipitate in erythroid precursors at bone marrow, causing membrane damage and cell death. Ineffective level of erythropoiesis is the main determinant of development anemia, while peripheral hemolysis of mature red blood cells and the overall reduction in Hb synthesis are secondary. They have a moderate hemolytic anemia, maintaining Hb levels at 7g/dL without transfusion support.19

Some patient with systemic diseases are prone to have Recurrent Aphthous Ulceration (RAU). The aetiology of RAU is unknown. The major factors identified include heredity, hematologic deficiencies, and immunologic abnormalities. These patients tend to have an autoimmune reaction that produces oral aphthous ulceration. The diagnosis of RAU is made on clinical grounds. These presentations have been termed pseudoaphthae or, more recently, aphthous-like ulceration (ALU).8,10,11

The main management strategies for TI are transfusion therapy, iron chelation, splenectomy, modulation of gammaglobulin chain production, drugs according to recommendations, adequate hydration, not biting lips, not licking lips, not rubbing the lips, and maintaining oral hygiene.
stem cell transplantation, and recent clinical experimental trials on Minihepcidin Peptide or similar drugs (ACE-536, ACE-011) that increase differentiation or maturation developing red blood cells in the bone marrow. Even though transfusions are irregular needed for TI patients, this is very important treatment options in several situations. Blood transfusion in TI, besides increasing intestinal iron absorption due to chronic hemolysis, anemia causes excess iron in some people patients, who need iron chelation therapy for prevention of iron deposition in critical organs. In this case, the patient had undergone blood transfusion therapy since he was 1.5 years old. Patient also consume folic acid and feriprox everyday. The results of laboratory tests, ferritin levels in patient was very high. Deferiprone (Ferriprox) is an iron-chelating agent in oral tablet form. Deferiprone treatment is relatively safe for patients suffering from various hematologic and oncologic diseases for which RBC transfusion is needed during treatment.

Infections are a frequent complication of thalassemias arises both from a large spectrum of immunological abnormalities and from the exposure to infectious agents. The pathogenesis of thalassemia is based on ineffective erythropoiesis, hemolysis, and the tendency to increase iron absorption lead immune disfunction because 2 reasons. The first reason causes monocytes/macrophages compartment become hyperplasia and hyperactivity in phagocytes all damaged erythroid precursors and erythrocytes. Increased phagocytic activity is very likely to reduce the capacity of the phagocytic system to survive against the pathogens of microorganisms. The second reason, severe anemia is a risk factor for bacterial infection in thalassemia. In these cases, anemia itself represents other risk factors for infection.

Management of RAU using topical and systemic therapies is based on severity of symptoms and the frequency, size, and number of lesions. The goals of therapy are to decrease pain and ulcer size and number, to promote ulcer healing, and to reduce the frequency of ulcer recurrence. The patient in this case is treated using chlorine dioxide oral rinse for ulcer in labial mucous and recovery was achieved within 3 weeks follow-up. Chlorine dioxide (ClO2) is a free radical that is soluble in water and stable for a long period of time without exposure to light. Previous research has shown that ClO2 is effective antimicrobial agent against many species of bacteria. When existing in the mouth, ClO2 reacts with amino acids found in saliva, which are nutrients for bacteria and interrupt the growth of bacteria through the nutrient pathway. ClO2 can penetrate biofilm easily and increase antibacterial efficiency. ClO2 can help to create an oxygenrich environment in the oral cavity, limiting the growth of bacteria, especially anaerobic species. ClO2 is not only bactericidal, but it has also been proven to be fungicidal and viricidal.

Conclusion
Hematological disorders frequently affect soft tissues of mouth with diverse characteristics. Management of oral manifestation in Thalassemia patient need multidisciplinary approach to obtain successful therapy. Advances in the treatment options resulted in long-term disease-free survival and improved the patient’s quality of life.

Aknowledgment
None.

Conflict of Interest
The authors report no conflict of interest.

References