

CASE REPORT

Clinical Management of a Pediatric Patient Associated with Thalassemia Major: A Case Report

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ABSTRACT

Aim: To discuss a case of beta thalassemia major with its clinical intraoral, extraoral features and radiographic appearances and clinical management.

Background: Thalassemia, a hereditary hemolytic anemia disease is the most widely distributed genetic disorder. It occurs due to a defect in the globin chain of hemoglobin. The more severe form, beta thalassemia major, presents with a diverse spectrum of clinical features caused by anemia, bone marrow hyperplasia, and blood transfusions.

Case description: Here is a rare case presentation of a 4-year-old male patient suffering from thalassemia major, reported with pain and swelling in the upper front teeth region. The patient was under regular blood transfusion regimen. The patient was diagnosed with having severe early childhood caries and managed with full mouth rehabilitation.

Conclusion: Thalassemia is characterized by diverse oral and dental features, a dentist should have complete knowledge of the thalassemia and its complications to treat the dental conditions effectively.

Clinical significance: Caregivers of children suffering from systemic illness lacks the incentive to maintain oral health. As a result, children suffer from comorbid conditions such as early childhood caries. Furthermore, the systemic condition also affects their oral health where the necessity for early intervention and conservative management is indispensable.

Keywords: Beta thalassemia major, Clinical management, Extraoral features, Intraoral, Severe early childhood caries.

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INTRODUCTION

Thalassemia is among the most widely distributed genetic disorder to cause a major public health problem.

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β -thalassemia is severe hemolytic anemia occurring as a result of the deficient or absent synthesis of globin chain of HbA.¹ This disorder is associated with a remarkable clinical heterogeneity with striking differences in hematological manifestations among different ethnic groups. The molecular basis of thalassemia has been studied worldwide. More than 200 different mutations in the globin gene have been reported. It was also found that types of mutations are ethnic specific.² The condition was first described by Cooley and Lee in 1925, and appeared in an article entitled "A series of cases of splenomegaly in children with anemia and peculiar bony changes".³ The term thalassemia was first used by Wipple and Bradford in 1932, derived from Greek word "thlassa" meaning sea and "hemia" meaning blood.²

Beta thalassemia is considered to be a major public health issue, as well as being a life threatening condition characterized by severe anemia, hepatosplenomegaly, growth retardation, endocrine dysfunction, cardiac failure, and skeletal changes. The hypertrophy and expansion of erythroid marrow is reflected at the skeletal level, and especially in the facial bone.⁴

Oral health of children suffering from thalassemia major is reported to be poor by most of the researchers. As low priority is given to the oral health status by the masses in the country in general, this negligence might be compounded for children already suffering from a life-threatening systemic disease because the parents might focus on the medical procedures required to overcome this disease during early childhood. So, this poor oral health, in turn, leads to further deterioration of systemic health in these children.⁵⁻⁸

CASE REPORT

A 4-year-old male patient visited the department of pedodontics with the chief complaint of pain in the left lower back teeth region for 15 days (Fig. 1). It was not associated with swelling. The patient was diagnosed with thalassemia major 3 years back and had been undergoing blood transfusion every month since then. Family history revealed the presence of thalassemia minor in both of his parents. The patient also gave a history of trauma one year back. The patient was poorly built and nourished with short stature. Extraoral examination of



Fig 1: A 4-year-old male beta thalassemia major patient

the patient revealed frontal bossing, malar prominence, with no maxillary proclination. Intraoral examination demonstrated generalized yellowish discoloration of teeth, carious left mandibular second molar, discolored maxillary left central incisor and localized gingival recession with upper central incisors. There was also initial occlusal caries in maxillary first molars, smooth surface caries in the right canine and proximal caries in the mandibular canines and first molars (Figs 2 and 3).

The patient was diagnosed to be a case of severe early childhood caries. Further, there was mucosal pallor and yellowish tinge at the junction of the hard and soft palate.

The hematologic investigation revealed microcytic hypochromic and normocytic normochromic anemia with features of mild chronic hemolysis which was consistent with the thalassemia. Later Hb electrophoresis was done which confirmed the diagnosis of beta thalassemia. The patient was regularly transfused with 1 unit of packed red blood cell every month.

Dental treatment was done on the following day of blood transfusion appointment. Full mouth rehabilitation was done with the endodontic treatment of left lower second molar and maxillary central incisors and glass ionomer cement restoration of all other carious lesions maintaining proper isolation. Stainless steel crown was placed following endodontic treatment of the lower molar. Considering the periodontal status of the maxillary left central incisor and the age of the patient, bleaching pro-

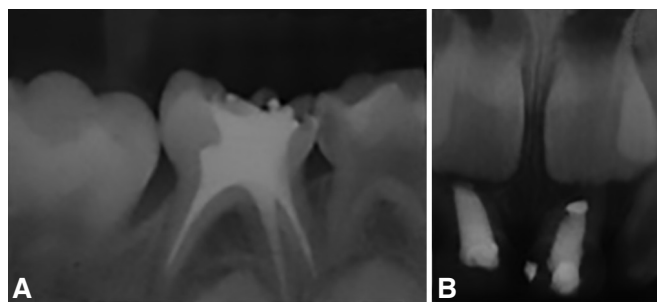
cedure to esthetically rehabilitate was not done (Figs 4 and 5).

DISCUSSION

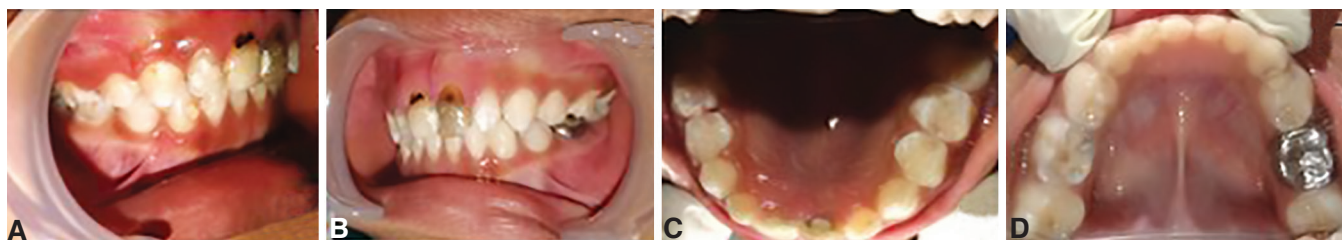
Thalassemias are globally widespread. The highest frequencies are observed in South-East Asia and sub-Saharan Africa.⁹ In India prevalence of β -thalassemia is 3 to 4% with an estimate of around 8,000–10,000 new births with the major disease each year.¹⁰ Thalassemia, one of the hemoglobinopathies often presents with a diverse clinical picture which may cause a diagnostic dilemma. Bone marrow hyperplasia and enlargement contributes to peculiar orodental manifestations.¹¹ Bone marrow hyperplasia or enlargement plays the main role in causing peculiar ‘chipmunk facies’ which is not distinguishable in our case, though frontal bossing and malar prominence can be noted.¹²

Anemia is the typical feature of thalassemia which results from the synthesis of abnormal hemoglobin. Microcytic hypochromic anemia subtype is more frequently observed in thalassemia major patients as early as 1st year of life as the fetal hemoglobin formation stops after 3 to 4 months.¹³ The same subtype of anemia was observed in our case. Fragile RBCs which are produced in thalassemia further results in increased hematopoiesis causing bone marrow hyperplasia.

The main therapeutic approach of thalassemia major is by transfusion of the blood every 2–3 weeks. Frequent transfusions result in iron overload which further causes secondary hemochromatosis, with the involvement of



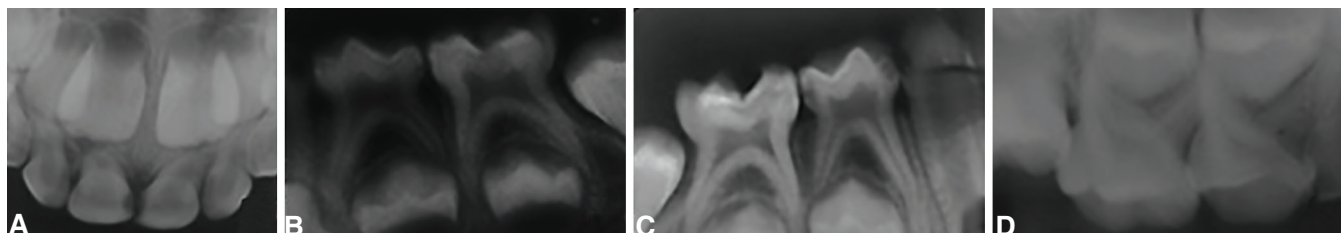
Figs 3A and B: Postoperative periapical radiographs: (A) Mandibular left quadrant; (B) Maxillary anterior



Figs 2A to D: Postoperative intraoral view: (A) Right lateral; (B) Left lateral; (C) Maxillary occlusal; (D) Mandibular occlusal



Figs 4A to C: Preoperative intraoral view: (A) Anterior; (B) Maxillary occlusal; (C) Mandibular occlusal



Figs 5A to D: Preoperative periapical radiographs: (A) Maxillary anterior; (B) mandibular right quadrant; (C) mandibular left quadrant; (D) maxillary left quadrant

heart, liver and endocrine glands.¹⁴ Yellowish tinge of oral mucosa as observed in our case is associated with thalassemia, caused by bilirubin formed from degradation of RBCs.¹⁵ Mucosal pallor and atrophic glossitis are also evident due to anemia, as in our case anemia was evident. Hyposalivation and inflammation of salivary gland also found in a thalassaemic individual due to iron deposition, which was not evident in the above case.

Bone marrow hyperplasia caused due to chronic anemia gives rise peculiar radiographic feature of thalassemia.¹⁶ Radiographically thalassemia characterized this enlargement is explained by the fact that when ineffective erythropoiesis damages the RBC membrane leading to severe anemia, the body responds by increasing the production of RBCs, consequently causing expansion of the bone marrow up to 15–30 times the normal amount.^{17,18} The findings are thin cortex, spiky shaped short root, faint lamina dura, and absence of inferior alveolar canals.

An important finding in our case was the gingival recession in the maxillary centrals. Akcali et al. hypothesized that thalassemia major may cause increased levels of inflammatory cytokines in biofluids, particularly when there is accompanying gingival inflammation, which in turn may modify the clinical signs of both chronic diseases.¹⁹ Altered bone metabolism, such as osteopenia and osteoporosis, is the major cause of morbidity in thalassemia major.²⁰ Receptor activator of nuclear factor- κ B (RANK), RANK ligand (RANKL), and osteoprotegerin (OPG) are members of the tumor necrosis factor (TNF) receptor superfamily, and together with various cytokines, they play a central role in bone remodeling.²¹ Alterations in the RANK/RANKL/OPG system are important in the impaired bone turnover

in patients with TM through complex mechanisms involving chronic anemia, iron toxicity, and endocrine complications.²⁰

The treatment for beta major thalassemia is performed routinely especially when patients experience decreasing hemoglobin level. Therefore, the dental treatment should be performed by paying attention to the hematologic profile of the patient and all dental treatments should be planned with the hematologist, which was followed in our case. The dental treatment was performed in a short time as possible and was done after the patient received a blood transfusion. No dental treatment was done when the hemoglobin level was less than 100 g/L. Antibiotic administration was done to the patient to treat chronic periapical infection in relation to the upper left anterior teeth. Oral hygiene measures were critically undertaken for this patient to avoid the spread of any infection. Pulp-ectomy procedure was performed for endodontically involved left lower primary second molar and upper primary central incisors with local anesthesia administration and maintaining proper isolation.

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