International Journal of Current Research and Review
DOI: https://doi.org/10.31782/IJCRR.2021.SP291

Scopus

Case Report

IJCRR
Section: Healthcare
ISI Impact Factor (2019-20): 1.628
IC Value (2019): 90.81
SJIF (2020) = 7.893

Oral Rehabilitation of Beta Thalassemia Patient Treated Chair-side - A Case Report

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ABSTRACT

Introduction: Thalassemia can be classified into alpha and beta depending on the reduced synthesis of alpha or beta chains present in hemoglobin (Hb),1 that results in reduced hemoglobin in body than normal.

Aims: Aims at signifing the importance of timely dental treatment in Beta thalassemia patients.

Case Report: Here we present a known case of beta thalassemia minor requiring a full mouth rehabilitation treatment performed dental chair-side.

Discussion: Thalassemia minor patients inherit only a single beta- globin mutation and have a mild anemia.2 The evaluated predominance of β-thalassemia minor (carriers) in India is 3-8%, which transforms to 35-45 million carriers in 1.3 billion diverse population with culturally and linguistically multi-ethnic people.3.

Conclusion: Several ethnic groups have a much higher prevalence of 4–17%.4 The Beta thalassemia minor cases usually go undetected and are often observed in tests for dental treatment or post dental treatment.

Key Words: Beta Thalassemia, Dental Chair, Oral rehabilitation, Hereditary, Red blood cells, Autosomal

INTRODUCTION

Thalassemia is a hereditary disease with at least one parent as a carrier for the disease, caused by either a deletion of certain key gene fragments or a genetic mutation. Thalassemia was first recognized by Dr. Thomas Cooley (1925), as an autosomal recessive blood disorder.[5] Hemoglobin consists of two proteins, an alpha, and a beta associated with the oxygen-carrying component of the red blood cells.

α and β-thalassemia are common throughout the world, among the various Hemoglobinopathies. The carriers of this genetic trait in about 7% of the global population. The Indian subcontinent is known to have 2-3% of the general population to be β-thalassemia carriers.[6] In India, 3.7% of its total population is a thalassemia carrier with 50,000 new cases per year.

Point mutations in the beta-globin gene is a cause for beta Thalassemia. There are two different globin genes, alpha and beta, carried on chromosome 15 and 11 respectively. On the basis of the mutation of beta - gene, its divided into three categories.[6] The less production of beta chains is due to a heterozygous mutation resulting in beta-thalassemia minor, which is mild and usually asymptomatic. A homozygous mutation of the beta-globin gene, leading to total absence of beta chains is the cause of beta thalassemia major. It expressed clinically as jaundice, endocrine abnormalities, growth retardation, hepatosplenomegaly, and severe anemia requiring repeated blood transfusions life-long. Beta-thalassemia intermedia is the third type presenting with mild to moderate clinical symptoms in comparison to the other two types.[1]

Although the mutations in β-thalassemia minor may decrease synthesis of the affected globin chain, producing hypochromic and microcytic RBCs, the anemia is usually modest with relatively little accumulation of the unaffected globin. Therefore morbidity associated with chronic hemolysis and ineffective erythropoiesis is rarely encountered.[2]

Beta thalassemia shows oral manifestation - prominent frontal bossing and zygomatic arches, dental, and skeletal maloc-

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ISSN: 2231-2196 (Print) **ISSN:** 0975-5241 (Online)

Received: 26.12.2020 **Revised:** 24.01.2021 **Accepted:** 24.02.2021 **Published:** 10.03.2021

clusion with overgrowth of the maxillary bone, along with delayed development of teeth. Children with Thalassemia minor do not show any specific oro-facial traits.[7]

Thus, based on these findings we present a case report of a patient with beta thalassemia minor treated on dental chair for oral rehabilitation.

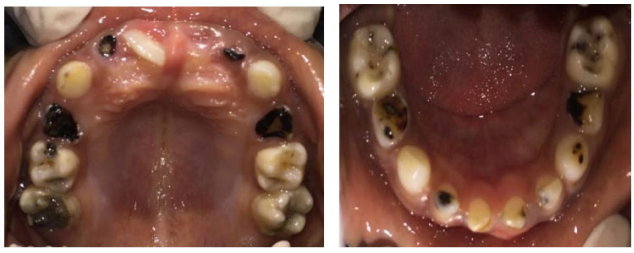
Case Presentation

A Male child aged 8 years reported to the Department of Pediatrics and Preventive dentistry with the chief complaint of pain in the upper right back region of the jaw since 2 months. The pain is intermittent which aggravates on chewing and persists till medication is administered, which was prescribed by general dentist (Ibuprofen).

The child was detected with Beta thalassemia during the 1st trimester of pregnancy, by a thalassemia screening test using Chorionic villus. The Mother has a history of beta thalassemia along with the older sibling of the child.

Child is on medication for same (Folic acid - 5mg). These is no history of blood transfusions done with a consistent Haemoglobin of 7 - 10gm/ dl. History of adenoid hypertrophy was detected 2 years ago.

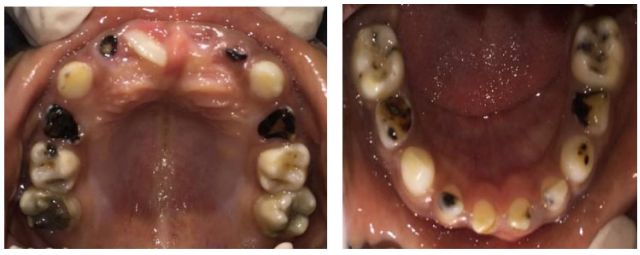
Patient presented oro-facial manifestation of prominent frontal bossing and zygomatic arches, flat nasal bridge, along with flared nostrils, dental malocclusion with delayed eruption of teeth.(Picture 1)



Maxillary arch Mandibular arch

Picture 1: Extra oral image.

On intra oral examination multiple carious teeth were observed. (Picture 2)



Maxillary arch Mandibular arch

Picture 2: Pre Operative Intra oral images.

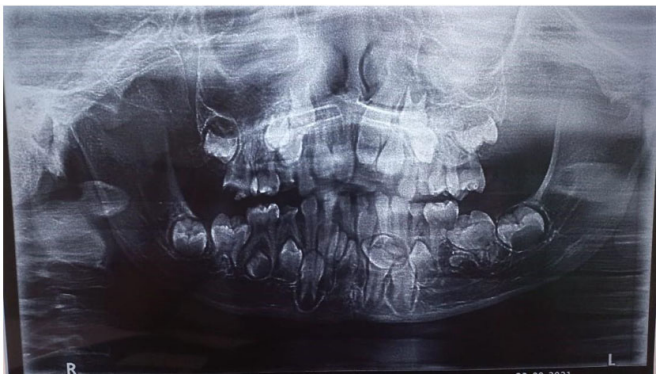
Investigations

Complete blood count (CBC) which revealed hemoglobin of 10 mm hg An orthopantamogram (OPG) was advised .

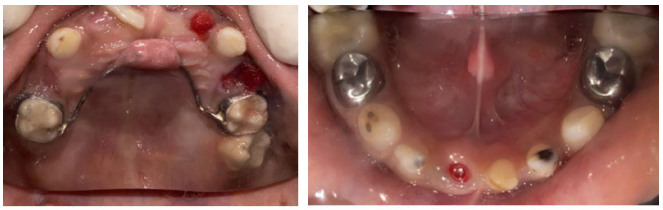
Treatment plan

On the basis of the OPG (Picture 3), the treatment plan was formulated, which was done in an isolated chamber with use of personal protective wear - Extraction of 16 was done due to pulpal involvement and less than 1/3rd root development as observed on intra oral periapical radiograph, followed by 54, 52, 62, 64, 71. Post extraction absorbable gelatin sponge (AbGel - Healthium Medtech) was used to control bleeding from the extraction sockets.

Restoration of 55,75,85,26; Pulpectomy followed by Stainless steel crown of 84; restoration followed by Stainless steel crown of 74 (Hall technique) done under rubber dam. Space maintainer for the maxillary arch (Nance's Palatal space maintainer).(Picture 4) The above treatment was carried out in two appointments on the dental chair.



Picture 3: Orthopantamogram (OPG).



Maxillary arch Mandibular arch

Picture 4: Post operative Intra oral images.

DISCUSSION

β-Thalassemia is the most prevalent variety of thalassemia, approximately 3% of the total world population is carrying the β-thalassemia gene. The March of Dimes Global Report on Birth Defects has estimated that the prevalence of pathological hemoglobinopathies in India is 1.2 per 1000 live births. [8,9,10]

The etiology of beta thalassemia is due to point mutations, more rarely, elimination in the beta globin gene present on chromosome 11, causing either absent (beta 0) or less (beta+) production. Diagnosis is based on hematologic and molecular genetic testing. Clinical diagnosis of the genetic disease is often confirmed by molecular detection of the mutational changes in the globin genes.[7]

Main clinical featuresof beta thalassemia are varied from gallstones, increased predisposition to thrombosis, painful leg ulcers, hypertrophy of erythroid marrow with medullary and extramedullary hematopoiesis and its complications which include osteoporosis, masses of erythropoietic tissue that primarily affect the liver, spleen, lymph nodes, chest and spine, and bone deformities and typical facial changes. [9]

Major changes seen in the facial region in thalassemia patients include the prominence of zygomatic bones and maxillary enlargement due to erythroid hyperplasia that is accompanied by depression of the nasal bridge. All of these changes may cause characteristic "Chipmunk or Rodent facies." Other dental defects include forward drifting of maxillary incisors, spacing between the teeth, anteriorly located open bite, protruding maxilla, malocclusions, including macroglossia and atrophic glossitis, pallor mucosa. Radiographically, absence of inferior alveolar canal, short roots, and taurodontism are not uncommon findings. [11]

Facial features result from extra-medullary hematopoiesis and compensatory growth of the bone marrow.[12] The observed maxillary enlargement can cause structural changes in the oral cavity such as teeth protrusion, spacing, occlusal deep bite, open bite and different degrees of malocclusion that predispose to dental problems.[13] High degrees of periodontal diseases and dental caries have been extensively reported in thalassemia patients.[14]

β-Thalassemia patients are seen to have a higher risk of developing dental caries as well as periodontal diseases. This may be due to chronicity of this disease resulting in neglect of their oral health and hygiene. [12]

In addition, patients may suffer from other oral conditions due to neglect of oral hygiene, increased level of serum ferritin, iron overload and the variations in salivary biochemical constituents.[15] The child's oral health has an impact on eating, talking, laughing and appearance. Pain that results from oral health problems may negatively affect the child's daily life.[16]

CONCLUSION

Thalassemia minor is clinically asymptomatic and generally goes un-noticed as it does not show any clinical features but the present case did show clinical features, that, besides a known history both family and personal, was helpful in coming to a treatment plan and excecution of the treatment plan.

The case report aims at signifying the importance of timely dental treatment in Beta thalassemia patients. Educating the parents or guardians after the oral hygiene and regular dental check up.

ACKNOWLEDGEMENT

I acknowledge the scholars whose articles are included in references to this manuscript. I am also thankful to the authors/editors/publishers of those articles and journals from where the literature for this article has been reviewed. I am extremely thankful to the editorial board of "International Journal of Current Research and Review" who have helped in the publication of this manuscript.

Source of Funding: NIL

Conflict of Interest: NIL

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