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ORIGINAL ARTICLE

Mandibular analysis of Beta thalassemia patients from hematological institutes of Karachi Pakistan.

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ABSTRACT… Objective: To study the mandibular changes in the β-thalassemia major patient. **Study Design:** Cross sectional study. **Setting:** Husaini Blood Bank and Institute of Hematological Diseases Karachi, Department of Oral and Maxillofacial Surgery, and Department of Orthodontics, Baqai Dental College Baqai Medical University, Karachi. **Period:** January 2023-December 2023. **Methods:** 50 Orthopanthogram (OPG) radiographs of beta-thalassemia major patient were taken, analyzed and compared to check any variation from normal. Fifty diagnosed β-thalassemia major patients were voluntarily selected through convenience sampling. The study was carried out in collaboration of Husaini Institute of Blood Diseases Karachi, National institute of Oral Diseases Karachi Pakistan and, Department of Oral and Maxillofacial Surgery and department of orthodontics Baqai Dental College Baqai Medical University. **Results:** 18 (36%) patient's alteration of trabecular pattern are present and in 32 (64%) patient alteration of trabecular pattern absent. Similarly, in 16 (32%) patients have short spiky roots are present and 34 (68%) patients short spiky roots absent. **Conclusion:** Effect of anemia is not so profound in mandible of thalassemia patients from population of Karachi.

Key words: Karachi, Mandibular, Oral, Pakistan, Thalassemia.

INTRODUCTION

Hemoglobin is either partially or completely absent from the blood due to the autosomal recessive blood illness known as thalassemia, which can be passed down through a single gene (-thalassemia) or many genes (-thalassemia).[1,2,3] The Mediterranean region experiences it frequently.[4] Two -globin chains and two -globin chains, grouped in a hetro-tetramer, make up the four protein chains that make up hemoglobin.[5]

Defects in either the globin chain that formed the defective red blood cells, or both, are present in patients with thalassemia.[2]

The severity of the disease varies depending on the type of HB-gene mutations that cause -thalassemia at chromosomal No. 11.

There are three subclasses according to severity. Thalassemia comes in three different severity levels: major, intermediate, and minor (the amount of -globin determines the severity of the condition).

However in each sub class tetramer do not form and they bind to the red blood cell membranes, causing damage to membrane.[6,7,8,9,10,11]

Two genes, HB1 and HB2, are implicated in -thalassemia and are inherited. This results in an excess of globin chains in adults and excess globin chains in newborns. The extra -chains combine to generate unstable tetramers that exhibit aberrant oxygen dissociation curves.[2,12,13] In general, hemoglobin is formed of and -chains; however, and chains make up only around 3% of adult hemoglobin. The creation of - chains is also impacted by mutations.[13] The general symptoms of thalassemic patients are caused by a total or partial loss of globin chain production,

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which has major effects on their bodies and has been thoroughly discussed by numerous workers.[2,3,7,8,14,15] Additionally, B-thalassemia is to blame for a variety of symptoms and consequences that affect patients' various organs to varying degrees.[1,16,17,18,19,20,21,22,23,24,25]

In β -thalassemia, the oral and maxillofacial manifestation has been reported in several reports.[14,16,17,18,22,24,26] In most of the part of the world Orthopanthogram (OPG) studies has been done by several workers.[13,21,22,24,27,28,29]

It takes a year for radiological changes to become visible. Large bone marrow gaps are one of the most significant radiographic characteristics of thalassemia.

The reason for this enlargement is that the body responds to severe anemia Whereas in the present study patients were also 50 with mean age of 20.1years with 32% spiky roots, closer to Babu and Amitha (2014)[28] Ash and Steiner (2020)[32] in Wheeler's Dental anatomy, physiology and occlusion book described teeth root development pattern in normal children and it is13-14 mm long in first and second molars.

Anemia caused by inefficient erythropoiesis by increasing the synthesis of red blood cells. This causes the bone marrow to enlarge by a factor of 15–30 times more than it would normally.

The radiograph of the skull reveals the expanded diploid space and vertical rows of trabeculae that give the appearance of hair on end.[21] The classical Chipmunk facies, which includes a depressed cranial vault, frontal bossing, maxillary expansion, a retracted upper lip and saddle nose, a yellowish tinge at the meeting point of the hard and soft palate, yellow-tinged fingernails, and spaces with widened trabeculae, is also attributed to bone marrow expansion.[24] Both angular and linear measurements showed that the growth direction was clearly vertical.

The most prominent dental abnormalities in thalassemic patients were incisor proclination, substantial over eruption, and increased over jet.

The measurements of the soft tissues revealed a markedly convex lower face and pronounced upper and lower lips. Since there has been no research on the radiological (OPG) orthopanthogram aspect of thalassemic patients in Pakistan, the current study has been conducted.

METHODS

Present study is carried out in the Husaini blood bank and Institute of Hematological diseases Karachi, National institute of Oral diseases Karachi and, Karachi and Department of orthodontics and Department of Oral and Maxillofacial Surgery Baqai Dental College Baqai Medical University and Department of Orthodontics during the time period of January 2023-December 2023 after taking approval from the IRB of Baqai University (Letter no.BDC/ERB/2023/030).

Sample Population

Since the rarity of disease with only prevalence rate of 5-7% and limited registered patients across Karachi, all 50 registered patients were included in the study population.[30]

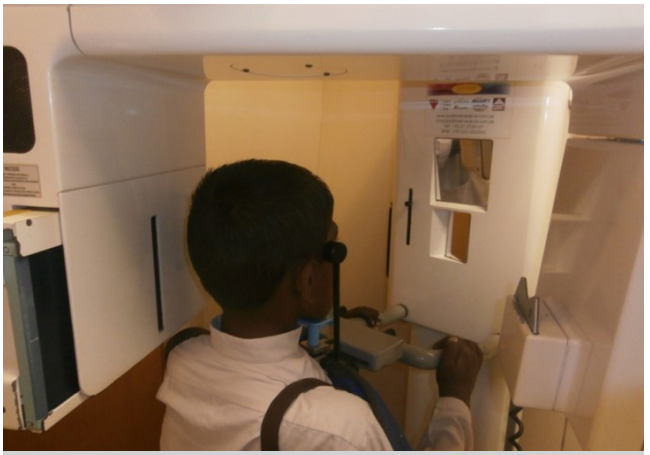
Methods for Collecting Data

From among the patients registered for treatment fifty diagnosed β – thalassemia major patients were selected through convenience sampling technique between the age range of 5-35 years. Exclusion criteria included patients under treatment or those who did not consent. Patients were approached by the investigators and their consent and ascent in cases of minors was obtained by the investigators prior to enrolling in the study. After that their orthopantamotomogram was taken to assess the dentoalevolar changes from norms.

Radiographic Procedure and Interpretation

a. Requirements

1) Room with subdue ambient illumination;
2) Radiographic viewer;
3) Magnifying lens; 4) Orthopanthogram (OPG)
5) All the processed radiographs.



Pic (A)

b. Positioning for OPG:
According to the setup of the panoramic machine described by Langland et al. (1982) [31], the patient was positioned correctly.
a) The patient was made to sit on the unit chair, which had a height-adjustable knob that was pressed.
b) The operation of the equipment was described to the patients. c) They were told to take their spectacles and any metallic objects off of their head and neck. d) The patient's jacket, jumper and other bulky items were taken off so that there was enough room between the bottom of the cassette holder and the patient's shoulder.
e) The exposure parameters were changed on the control panel.
f) The patient was asked to sit upright with a straight back and a slightly forward-extended neck. g) A disposable bite block envelope was used to cover the bite block.

The patient was instructed to purse her lips and press her tongue firmly against her palate. i) Adjustments were made to the temple supports.
j) The patient's head was tipped downward so that the line from the tragus to the ala is 50 degrees down and forward.
k) By having the patient bite with his central incisors (upper and lower teeth), the patient's mid-sagittal plane was placed in the center of the x-ray unit's focal trough.

RESULTS

Patients who were enrolled in the study met the requirements for inclusion and exclusion as well as for statistical analysis of the data using IBM SPSS version 23. Around 50 patients were included in the study who were registered but waiting for treatment. There were 28 male and 22 female patients. The majority of the patients belonged to the age group 5-35 years.

Table-I shows that only 18 (36%) had alteration of trabecular pattern present on the other hand, 37 (74%) had normal pattern. Similarly, in 16 (32%) patients short spiky roots were seen while 34 (68%) patients had normal root morphology.

Variables	Frequency (n=50)	Percentage
Alteration of trabecular pattern	18	36%
Short spiky roots	16	32%

Table-I. Alteration of trabecular pattern and Short Spiky Roots

DISCUSSION

In the current investigation, radiographic examinations of 50 - thalassemic individuals were done; 18 (36%) of them had altered trabeculae, whereas 32 (64%) had normal trabeculae.

While Poyton and Davey (1968) reported 87.5% alteration of trabeculae pattern, Kalpan et al. (1964) reported 86%, and Babu and Amitha (2014) reported 84% of patients have alterations in trabecular pattern, the present study reported only 36% of the cases, which is very low and is a positive indicator of proper and consistent patient care.[32,28]

Bone marrow hyperplasia mainly responsible for these changes and thus causing thinning of cortical borders. Out of 50 patients, short spiky roots of teeth were found in 16(32%) patients and normal roots were present in 34(68%) patients. Patil (2006)[16] and Parkin (1968)[1] reported 12% spiky roots. Patil (2006)[16] in her study take the thalassemic patients of age rang 3-28 years and their mean age was 8.5 years. In most of the patients, roots were not fully developed.

Where as in the present study age range was 6 to 35 years, and mean age 20.1 years and majority

of the patients have fully developed roots. 68% normal roots and 32% spiky roots.

Patil (2005)[16] and Parkin (1968)[1] reported 12% spiky roots which is less than half reported in the present study. Babu and Amitha (2014)[28] carried out radiological study of 50 children suffering in β- thalassemia with mean age of 16 years and reported 34% short spiky roots.

Whereas it is 9-12 mm in thalassemic children growth rate in thalassemic child. It is slow than normal children, which ultimately affect tooth and its root, growth in size and dimension in thalassemia major. Poyton and Davey 1968)[32] and Hazza and Al-Jamal (2006) have also reported teeth with short spiky roots.

CONCLUSION

Thalassemia patients from Karachi, Pakistan, do not have severe anemia symptoms. On evaluating the X-Rays of the patients showed spiky roots and trabecular changes.

RECOMMENDATION

1. Consanguineous and intercommunity marriages are discouraged or outlawed (This is a particularly challenging task in Pakistan, where the population is founded on a closed family and community system.

However, it can be accomplished via public awareness campaigns through any channel (electronic, social, print, lectures, or seminars), as well as through any available media.

2. Although the Sindh Assembly and Pakistan's National Assembly have already enacted bills requiring thalassemia testing prior to marriage, this need must be enforced. Only legislation implementation is a concern right now.
3. More institutes for thalassemia and bone transplantation should be built by both the private and public sectors with the aid of international donor organizations or through public donations, where free or low-cost treatment is provided to patients.
4. The Pakistani government should take the necessary steps to set up free annual dental exams and dental care for thalassemia patients in both public and private hospitals.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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2	Syed Mahmood Haider	Supervisor of project main idea.	
3	Kashif Ikram	Critical review, Proof reading.	
4	Muhammad Uzair	Data collection, Data analysis.	
5	Sana Ahmed	Critical review, Data analysis, Data collection, Writing.	
6	Syed Azhar Matloob	Critical review, Data collection