Orofacial bone complications in thalassemic children associated with cephalometric evaluation.

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Abstract

Introduction: Beta Thalassemia (BT) disorders are characterized by a genetic abnormality in the compound of beta-globin chains, one of the most common hereditary diseases in Turkey and worldwide. Some of the characteristic orofacial complication of BT is associated with bone structure.

Material and method: Thalassemic and control groups selected retrospectively from the archives of Dortcelik Pediatric Hospital, Bursa. Cephalometric measurements were used in the current study. All statistical analyses were conducted using student’s-test and Mann-Whitney U test.

Results: Significant findings were shorter SNB angles, greater ANB angles, greater condylion to point A sizes, protrusive upper lips, shorter interincisal angles, greater value of wits, shorter interincisal angle, greater U1-NA, shorter anterior face height (NaMe), shorter posterior face height (SGo) are significantly in thalassemic patients (n: 26). All measurements have indicated affected orofacial structure.

Conclusions: Orofacial complication due to bone structure development in thalassemic patients is pronounced. Distinct findings are treated by orthodontics. When orofacial intervention is applied, the complications were evaluated. All treatments must be controlled by a hematologist.

Keywords: Thalassemia, Developmental toxicology, Orofacial complications.

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Introduction

Beta-thalassemia major (BT) is considered one of the most prevalent genetic anomalies in the world, including in countries like Turkey [1,2]. These are a group of hereditary blood disorders characterized by abnormalities in the composition of the beta chains of hemoglobin. The total annual Beta-thalassemia incidence of symptomatic individuals is estimated at 1 in 100,000 in worldwide and 1 in 10,000 people in the European Union [3].

The patients of BT usually come to medical practice within the initial years of life and require successive RBC transfusions to stay alive. The clinical feature of BT in serious findings include growth retardation, pallor, jaundice, poor musculature, genu valgum, hepatosplenomegaly, leg ulcers, extramedullary hematopoiensis, and skeletal deformities caused by enlargement of the bone marrow.

The deformities involve changes in the long bones of the legs and distinctive craniofacial changes (bossing of the skull, prominent malar eminence, depression of the bridge of the nose, tendency to a mongoloid slant of the eye, and hypertrophy of the maxillae, which tends to expose the upper teeth) [4-7]. The distinctive features of orofacial findings of BT are prominent cheekbones and protrusive premaxilla. Cephalometric analysis reveals dilatation of the diplopic space, subperiostal bone grows, partialy obliterated maxillary sinus. The findings are due to extra medullar erythroid hyperplasia [8-11].

According to our knowledge, scarcely any detailed studies on craniofacial measurements and deformities in thalassemic population have been published. We aimed to perform cephalometric analysis of patients with thalassemic patients consistent with the most widely-known orofacial clinical manifestations.

Materials and Methods

Records of talasemia patients and heath individuals (control group) were selected retrospectively from the archives of Dortcelik Pediatric Hospital. Selection criteria included no previous orthodontic treatment and having cephalometric films and records for talasemia, age and sex mismatch. A control group was selected from records of Class I malocclusion patients with no systemic disease who came to the dentistry clinic of Dortcelik Pediatric Hospital.

Standardized lateral cephalograms of 26 talasemia patients (17 girls, 9 boys) and age and sex-matched 30 healthy individuals that had been taken with the same digital cephalometric imaging system (Blue X Imaging Srl, Assango, Italy) in a natural head position were selected. Cranial and dentofacial parameters were measured using a cephalometric analysis...
program (Nemoceph Imaging Cephalometric and Tracing Software S.L, Spain).

Figure 1: Cephalometric linear measurement.

Figure 2: Cephalometric angular measurement.

A total of 26 measurements were performed on the lateral cephalograms. Cephalometric landmarks were marked and digitized by one of the authors. Cephalometric reference points and planes used in the study are shown in Figures 1 and 2. Used in cephalometric landmarks are listed in Table 1 [12].

<table>
<thead>
<tr>
<th>Table 1: Cephalometric landmarks.</th>
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<tbody>
<tr>
<td>N</td>
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Statistical method

All statistical analyses were conducted using SPSS version 13.0 (Statistical Package for Social Sciences). Student’s-test and Mann-Whitney U test was used on between-group comparisons. All statistical data, p<0.05 was accepted as significant.

Results

Thalassemic patients (n:26) mean age is 14.07 ± 3.27 and control group mean age is 12.57 ± 2.76. Either group’s ages were the same. (p=0.068). Sex ratio of patients was M/F, 9 (34.6%)/ 17(65.4%), and control group was M/F 14 (46.7%)/ 16(53.3%). Either group was same for sex. (p=0.361).

Table 2: Comparison of the cephalometric variables.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Thalassemic Patients</th>
<th>Healthy Individuals</th>
<th>P</th>
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<tr>
<td></td>
<td>Mean</td>
<td>SD</td>
<td>Minimum</td>
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<tr>
<td>ANB (*)</td>
<td>5.43</td>
<td>2.41</td>
<td>-1.5</td>
</tr>
<tr>
<td>SNA (*)</td>
<td>79.5</td>
<td>3.45</td>
<td>70.6</td>
</tr>
<tr>
<td>SNB (*)</td>
<td>74.07</td>
<td>3.48</td>
<td>65.4</td>
</tr>
<tr>
<td>GoGn-SN</td>
<td>34.93</td>
<td>6.66</td>
<td>15</td>
</tr>
<tr>
<td>Anterior face height (NaMe)</td>
<td>106.7</td>
<td>8.38</td>
<td>90</td>
</tr>
<tr>
<td>Posterior face height (SGo)</td>
<td>65.69</td>
<td>5.37</td>
<td>54</td>
</tr>
<tr>
<td>P-A face height (%)</td>
<td>61.69</td>
<td>3.93</td>
<td>54</td>
</tr>
<tr>
<td>Lower anterior facial height</td>
<td>61.96</td>
<td>6.72</td>
<td>48</td>
</tr>
<tr>
<td>Ramus height</td>
<td>38.38</td>
<td>3.99</td>
<td>32</td>
</tr>
<tr>
<td>Mandibular body</td>
<td>63.69</td>
<td>5.12</td>
<td>54</td>
</tr>
</tbody>
</table>
Cephalometric measurements and the relationship between patient and control groups were summarized in Table 2. SNB (p=0.001) angles were significantly shorter in the thalassemic group while ANB (p=0.0001) angles were significantly greater. Posterior face height (SGo) (p=0.025), and P-A face height (%) (p=0.033) were significantly shorter. Condylion to point A sizes were significantly greater. The upper lips were protrusive (p=0.013). The interincisal angles were significantly shorter (p=0.047). Value of wits were significantly greater (p=0.0001). Interincisal angles were significantly shorter (0.047). U1-NA measurements were significantly greater (p=0.016).

**Discussion**

Cephalometric characteristics of thalassaemic patients are very important for orofacial application such as orthodontics. Reports of orofacial complications of thalassaemic patients are inadequate in the literature. These complications are important for orthodontic and surgical procedures in the patients [13-15].

Significant greater ANB angles in thalassaemia major patients attributed to a short mandible. This finding was consistent with the literature [13-15]. The SNB measurements have shown mandibular regression and increase in the thickness of the soft tissue of the chin in thalassaemic kids. Short mandibles were reported in thalassaemic patients some of our findings were associated with a short mandible [13-16].

Shortness of SGo and P-A face height was founded. These findings were similar to literature [13,15].

The condylion to point A measurement and upper lip protrusion in thalassaemic patients show maxillary protrusion. These findings contribute to the typical appearance of thalassaemic patients [11,17]. Differently, Abu Alhaija et al. and Amini claimed normal Maxilla length of thalassaemic patients [12,13]. The porgnathism was attributed by a short mandible. This finding was consistent with the literature [13]. Interincisal angles and wits associated with the biting process are affected in thalassaemic patient. We came across similar findings in the literature, but not exact matches [7,13].

Orofacial abnormalities in these patients are important in three aspects. Firstly, hemoglobin values of these patients need to be kept at an optimal level (base line of hemoglobin level between 9 and 10 gr/dL). This application will reduce the occurrence orofacial complications [13,18,19]. Secondly, orofacial complications should be treated by an orthodontist. Cephalometric evaluation must be done during orthodontic treatment. Characteristic complications of thalassaemic patients are well known. This knowledge is necessary for any orofacial intervention. And finally, all treatment plans should be consulted to a hematologist.

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