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Review Article

Medical and Dental Implications of Patients with Beta Thalassaemia Major. Part 2: Orofacial and Dental Characteristics: A Review

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Abstract

Thalassaemia, one of the most common genetic disorders, often causes serious medical, social, and psychological problems. Beta thalassaemia major is a life-threatening disorder that presents with a vast variability in the systemic signs and symptoms. In addition, orofacial and dental tissues are also affected. The common orofacial features among thalassaemic patients include: frontal bossing, skeletal overgrowth with characteristic appearances known as chipmunk faces, upper lip retraction, protrusion of pre maxilla bone associated with alveolar enlargement that causes malocclusion in the dentition with the clinical appearance of protrusion, flaring, spacing of anterior teeth and anterior open bite. The oral mucosa appears pale or a lemon yellow colour due to deposition of bilirubin pigmentation and anaemia. Sometimes the gingival colour tends to be dark, caused by high ferritin level in the blood.

Current reports show a significant improvement in thalassaemia major patients' survival rates. With increased life expectancy, the need for improved oral healthcare is very important to ensure a high quality of life for this patient population.

This paper reviews the literatures and discusses briefly the dento-facial manifestations, radiographic features, dental caries, periodontal and soft tissue conditions related to beta thalassaemia major as well as dental management and considerations of thalassaemia patients.

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INTRODUCTION

The term Thalassaemia derives from the Greek word “thlassa” meaning sea and “haemia” meaning blood, and was first used by Wipple and Bradford in 1932 [1]. Beta thalassaemia major, exhibits the most severe clinical symptoms and includes marked orofacial deformities [2].

The common orofacial features among thalassaemic patients include frontal bossing, skeletal overgrowth with characteristic appearances known as chipmunk faces, upper lip retraction, protrusion of pre maxilla bone associated with alveolar enlargement that causes malocclusion in the dentition with the clinical appearance of protrusion, flaring, spacing of anterior teeth and anterior open bite. The oral mucosa appears pale or a lemon yellow colour due to deposition of bilirubin pigmentation and anaemia. Sometimes the gingival colour tends to be dark, caused by high ferritin level in the blood [3,4]. The chronic nature of thalassaemia may explain the level of oral disease; affected individuals are preoccupied with their life-threatening problems and neglecting basic preventive dental care. The higher caries level can be also attributed to several factors including: poor oral hygiene, improper dietary habits, bone deformities, malocclusion, higher rate of infections and iron overload in parotid glands, dry mouth and lack of dental knowledge in these patients. Therefore, the dental caries experience is usually higher in thalassaemic patients [5].

This paper reviews and discusses briefly the orofacial and dental characteristic as well as dental management and considerations of thalassaemia patient.

LITERATURE REVIEW

Dento-facial manifestations

The most common craniofacial deformity in the thalassaemic patient is Class II skeletal base relationship with short mandible, a reduced posterior facial height, and an increased anterior facial proportion [4]. In addition, it has been reported in the literature that the severity of the disease is closely related to the degree of cephalofacial deformities. The major facial discrepancies in thalassaemic patients include prominent frontal and parietal bones, sunken nose bridge, protruding zygomas and mongoloid slanting eyes, enlargement of maxilla and obliteration of maxillary sinus due to bony expansion, with depression of the bridge of the nose. These features will result in the characteristic facial appearance called “Chipmunk face” [3,6].

In addition, the dental arch morphology in thalassaemic patients differs from that of the unaffected population; this may include a narrower maxilla and a smaller incisor width for maxillary and mandibular arches [7]. The overdevelopment of the maxilla results in increased over jet, spacing, and protrusion of upper anterior teeth, as well severe over bite and open bite [8]. It was also found that the dental development in beta thalassaemic children is delayed by a mean of 1.01 years compared with normal children; this delay is consistent with significant general growth retardation in this group [4].

Radiographic features

The radiographic features in beta thalassaemia major patients

were extensively studied using intra oral periapical views of the posterior region, lateral cephalometric and panoramic radiographs [9].

Skull: Thickening of the calvarium of the skull is mostly observed in the frontal region [10]. One of the most significant radiographic indicators of beta thalassaemia is “hair on end” appearance. This radiographic appearance is due to the extreme thickening of diploe; the inner and the outer plates become poorly defined and the trabeculae between the plates become elongated, producing a bristle-like appearance on the surface of the skull [3].

Teeth and jaws: short and spike-shaped roots may be observed with the highest frequency in the mandibular first molars and central incisors, taurodontism and thin lamina dura may also be observed. The maxilla radiographs demonstrate sharply demarcated intermaxillary sutures, prominent anterior maxilla and small maxillary sinuses [11]. In the jaws, there is generalized rarefaction of the alveolar bone, thinning of cortical bone, and a “chicken-wire” appearance of enlarged marrow spaces and coarse trabeculation [12], as well as faint inferior alveolar canal, and thin cortex of mandible [13].

Dental caries in beta thalassaemia major patients

Dental caries is defined as a progressive, irreversible, microbial disease results in the demineralization of the inorganic constituents, and dissolution of the organic constituents, thereby leading to cavity formation [14]. It is a multifactorial disease in which there is interplay of three principal factors: the host, microflora, and substrate. In addition, time is considered the fourth factor which influences dental caries [15,16].

In addition, dental caries is affected by factors within the host, which may be related to the structure of dental enamel, immunologic response to cariogenic bacteria, and the composition of saliva. The factors of saliva that have been proven to be associated with increased risk of dental caries include salivary flow rate, buffer capacity and pH, oral clearance rate, immunoglobulin, and innate non-immunoglobulin factors [17,18].

Studies have shown that patients with thalassaemia have a higher rate of dental decay than the normal population. This may be because patients have difficulty accessing regular dental care, or may be reluctant to attend if they feel the dentist does not understand their condition. However, patients may also be more concerned with potentially serious medical complications of thalassaemia, leading to neglect in oral hygiene. In addition, the median saliva concentrations of phosphorous and IgA are significantly lower in such patients which makes them more prone to dental caries [19]. Al-Wahadni, et al., reported in their study that a higher risk of dental caries was found in the thalassaemic group compared to normal groups [20]. On the other hand, Scutellari et al., found similar incidence of dental caries in beta thalassaemic patients and normal controls. Lower concentrations of immunoglobulin A (IgA) in saliva of thalassaemic patients have been claimed to be related to higher rates of dental caries [21].

A study conducted in Jordan reported the prevalence and distribution of dental caries in thalassaemia major patients using

the DMFT index and found that the prevalence of dental caries in thalassaemia major patients was higher when compared with the normal counterparts [22].

Periodontal and soft tissue conditions related to beta thalassaemia major patients

In children with healthy gingival and periodontal conditions, the gingival margin is several millimetres coronal to the cemento-enamel junction (CEJ). The gingival sulcus may possibly be 0.5-3mm deep on a fully erupted tooth. In teenagers with a healthy periodontium the alveolar crest is situated between 0.4 - 1.9 mm apical to the CEJ [23]. There are numerous forms of periodontal disease, which can affect children and adolescents; based on the 1999 International Workshop for the Classification of Periodontal Diseases and Conditions (Armitage, 1999; Clerehugh et al 2004) these include: gingival diseases, chronic periodontitis and aggressive periodontitis in addition to a spectrum of conditions [24,25].

Little data is available on the association of periodontal disease in beta thalassaemia major patients [20]. A recent study conducted in Italy [26], assessed the prevalence of periodontal disease, orofacial changes and craniofacial abnormalities in patients with thalassaemia major. Clinical and radiographic examinations were carried out. The sample consisted of 100 patients affected by beta thalassaemia major and 98 patients with beta thalassaemia intermedia; each sample was compared to the respective control groups of healthy individuals. Patients were examined for plaque deposits, gingivitis, periodontitis using Löe and Silness plaque and gingival indices. The results showed a significantly increased level of periodontal attachment loss in older patients with beta thalassaemia. Another study [10], was conducted to assess the prevalence of periodontal disease, orofacial changes and craniofacial abnormalities in patients with thalassaemia major. The sample consisted of 54 patients; and a similar number of unaffected individuals matched by age and sex served as a control. The results conclude that poor oral hygiene and gingivitis were observed in thalassaemic patient compared to control group. Subsequently, a study was conducted to know of any association between increased severities of periodontal disease in thalassaemic patients. The study concluded that thalassaemia major was not associated with increased prevalence of gingivitis or periodontitis [20].

Dental management and considerations related to beta thalassaemia major patient

Dentists should be attentive to the complications of thalassaemia to ensure safe dental management to these patients. All dental treatments planned should be in liaison with the haematologist. Dental appointments should be kept as short as possible, to avoid fatiguing of the patient. Furthermore, all dental procedures should be performed closely after the patient has received blood transfusion [27]. Patients with beta thalassaemia should always be asked specifically about a history of splenectomy. A patient who has had splenectomy is at risk of massive infection following bacteraemia due to an immune defect. Antibiotic prophylaxis prior to dental treatment should be considered for those patients [28]. Patients with thalassaemia are also at an increased risk of viral infection such Hepatitis B, C

and HIV; as such, all members of the dental team should be aware of this and take appropriate history information from the patient as well as proper infection control precautions during dental treatment of these patient [28].

Most children with thalassaemia may be treated regularly, using local analgesia, supplemented with inhalation sedation if necessary. The use of general anaesthesia should be avoided if possible because of the difficulties caused by low Hb levels and cardiac insufficiency, and enlargement of the maxilla caused by bone marrow expansion, which may cause difficulties in intubation for induction of general anaesthesia [28]. Oral complications are common in the thalassaemic patient, however, painful swelling of the parotid gland and xerostomia caused by iron deposition in the serous cells and a sore or burning tongue related to the foliate deficiency, in particular, may occur [28].

CONCLUSIONS

The risk of oral diseases in thalassaemia patients remain high and prevention against oral diseases therefore is very important considering the higher life expectancy of these patients and the role of good oral health status has increased the quality of life. A unified approach to dental care is essential, including close liaison between haematologists and paediatricians. The dentist, especially the paediatric dentist, plays a vital role in educating such patients and parents regarding the prevention of dental caries, and the importance of maintaining good oral hygiene. These patients present with several medical conditions that are of great concern to the dentist such as anaemia, splenectomy, and increased risk of viral hepatitis. Oral and dental problems can be minimized through essential oral health programs, supported by local health authorities. It is important to investigate the healthcare system provided for children with thalassaemia; this includes dental appointments, dental follow-ups, and waiting lists. Focus on parental awareness programs, which stress the importance of maintaining oral health in children with thalassaemia can provide for better care for these patients.

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