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Dentistry

LOOKING BEYOND THE ORAL CAVITY – A BETA THALASSEMIA MAJOR CASE REPORT

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ABSTRACT Individuals classified as having special health care needs (SHCN) may be more susceptible to oral disease over their lifetime and may need specialised dental care because of their medical conditions or other constraint. Beta Thalassemia Major is a hereditary hemoglobinopathy that can take on different clinical expressions, with intense and multiple orofacial manifestations. Dental professionals must possess in-depth knowledge of the nature of this disorder and must work with a multidisciplinary approach involving many medical professionals. In the present case, a 3-year-old male with a beta thalassemia major was referred to Department of Paediatric and Preventive dentistry where definitive treatment to eliminate all dental foci of infection treatment was performed under general anaesthesia. Multiple tooth extraction was performed. Because of their clinical circumstances, these patients require specialised customized treatment, which makes managing and changing their behaviour essential to providing high-quality dental care.

KEYWORDS : Beta Thalassemia Major, child, special health care needs.

INTRODUCTION

The AAPD defines special health care needs as "any physical, developmental, mental, sensory, behavioral, cognitive, or emotional impairment or limiting condition that requires medical management, health care intervention, and/or use of specialized services or programs.[1]

Thalassemia's are common all over the world. Historically, the Mediterranean region, the Middle East, and Southeast Asia have had the greatest rates of β-thalassemia, whereas Northern Europe and North America have had the lowest rates. Cyprus, Sicily, Sardinia, and Greece have high frequencies of B-thalassemia (5–15%) in the Mediterranean region, but other parts of Europe have far lower prevalence. B-thalassemia is primarily observed in immigrant populations in the United Kingdom. The frequencies range from 2 to 5% in the majority of the Middle East and West Asian nations. The frequency of B-thalassemia in the Americas is also low, appearing to be primarily among immigrant populations, whereas in North Africa it varies from 2 to 9% in various nations.[2] The major condition affects an estimated 8,000–10,000 newborns in India each year, and the prevalence of β-thalassemia ranges from 3–4%.[3] Only roughly 200,000 people with thalassemia major are known to be alive and listed as receiving regular treatment worldwide, according to Thalassemia International Federation.[4]

Special Health Care Needs problems can be linked to children who have behavioural (anxiety, ADHD, autism spectrum disorder), congenital (trisomy21, congenital heart disease), developmental (cerebral palsy), or cognitive disorders. Moreover, systemic illnesses and intellectual disability are included in SHCN.[1]

β-thalassemia is severe haemolytic anaemia occurring as a result of the deficient or absent synthesis of globin chain of HbA leading to anaemia. This disorder is associated with a remarkable clinical heterogeneity with striking differences in haematological manifestations among different ethnic groups.[5] Beta-thalassemia major is inherited in an autosomal recessive manner. If both parents are known to be heterozygous for an HBB pathogenic variant, each sibling of an affected individual has at conception a 25% chance of being affected, a 50% chance of being a (typically) asymptomatic carrier, and a 25% chance of being unaffected and not a carrier.[6]

Kaur et al observed a higher incidence of dental caries in patients diagnosed with thalassemia when compared with normal children.[7] Therefore, these children's oral health continues to deteriorate requiring a more elaborate treatment as against minor restorations and prevention. This may be due to lack of awareness and because of being over burdened with treatment of the disease and its prominent symptoms.

The purpose of this case report is to highlight the significance of having a holistic approach towards dental treatment of a child with beta thalassemia major which is a part of special health care needs

Case Report

A 3-year-old child came to the department of pediatric and preventive dentistry with a chief complaint of pain in upper and lower right and left back region of jaw since 5 months. The patient was a known case of beta thalassemia major that was diagnosed at the age of 6 months and had been undergoing blood transfusion every month since then. Patient gave a history of multiple episodes of fever and diarrhoea resulting in his body subsequently becoming pale and fragile and yellow in colour at 6 months of age. On haemoglobin electrophoresis test, it was revealed that the child was suffering from beta thalassemia major. Patient had a history of Congenital Heart Disease (Patent Foramen Ovale). 2D echo reports showed that he has a left to right shunt in the atrium. Patient had a good biventricular function and hence the patient was not advised to undergo any heart surgery for the same. Family history revealed the presence of thalassemia minor in both of his parents and they had a consangious marriage.

Diet history revealed that patient was breast fed till 2.5 years of age. Patient was on a restricted diet to avoid iron rich food items in his diet.

Patient needed chemotherapy and bone marrow transplant, after genetic and immunocompatbility testing, father was selected to be the

donor with a 50% cross matching. Intraoral examination revealed deep proximal caries present with 51,54,55 61, 64, 65, 85; root pieces with 52,62,63 and deep occlusal caries with 84. (figure1,2) OPG was taken for the patient and assessed. (figure 3)

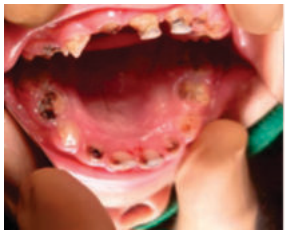

Figure 1

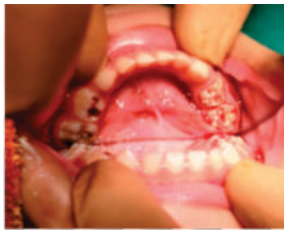

Figure 2

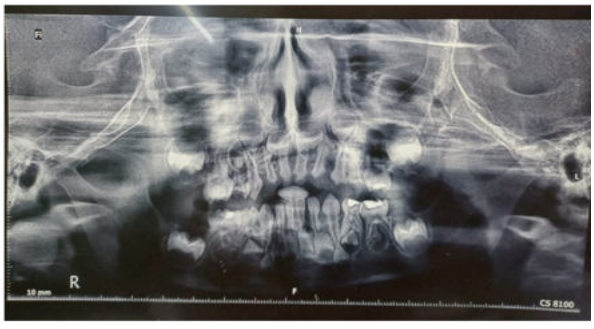

Figure 3

Due to multiple extractions, restoration and age of child, the patient was further evaluated for the case under general anaesthesia. Routine blood investigations were carried out which revealed that the patient had 9.6g/dl of Hb and o positive blood group. PA Chest and lateral nasopharynx Xray was taken. The necessary consents were obtained from paediatric haematologist, cardiologist, oncologist along with paediatrician and anaesthetist. Haematological investigations of the child after the recent blood transfusion; 2 days prior to the treatment revealed an Hb value of 11.7g/dl. Child was taken under G.A where extraction was preferred to endodontic therapy in view of avoiding bacteraemia and any spread of any infection systemically through oral cavity as post bone marrow transplant and chemotherapy child would be immunocompromised and prone to infections. L.A without adrenaline was used for extraction following which the bleeding was controlled with ab gel and 3-0 resorbable vicryl sutures were given to the patient. (figure 5,6). Child was kept in recovery and discharged next day. Follow up after 10 days showed an uneventful healing.

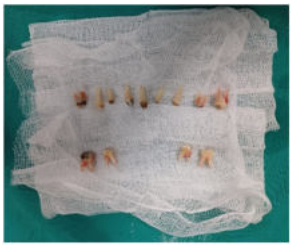

Figure 5

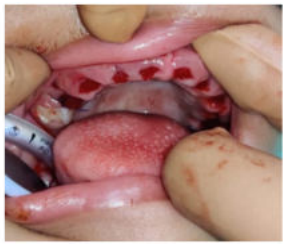

Figure 6

DISCUSSION

Beta Thalassemia is a blood disorder that reduces the production of hemoglobin, the iron-containing protein in red blood cells that carries oxygen to cells throughout the body this leads to a lack of oxygen in many parts of the body and causes a host of complications, including anemia. In children, beta thalassemia can significantly impact growth and development.[3]

One hemoglobinopathy, thalassemia, can have a variety of clinical presentations, making diagnosis difficult. One factor contributing to unusual orodental symptoms is bone marrow hyperplasia and enlargement, playing a major role in chipmunk facies, which is not distinguishable in our case, as the child was diagnosed early in his life with thalassemia and was started with blood transfusions and treatment. This also eliminated anaemia in our case, which is the typical feature of thalassemia.[5]

Blood transfusions raise levels of iron buildup, in addition to improving the prognosis for thalassemia major. Iron overload in beta thalassemia major individuals can be catastrophic if left untreated and cause multiple organ damage. Patients with thalassemia major experience an accumulation of iron in their gingiva, hepatic, cardiac, and endocrine organs. Iron accumulation can also lead to severe salivary gland oedema and inflammation as well as reduced saliva flow (xerostomia). Chelation therapy must therefore be used in order to eliminate the extra iron. Thus, the patient was counselled to abstain from iron-rich foods and to begin using the chelating agent: Deferoxamine medicine(defrijet) following dental rehabilitation treatment.[8]

Dental care planning is an important aspect for patients with beta thalassemia major. Multidisciplinary teamwork is essential for providing the dental treatment possible for thalassemia patients. To ensure that risk is kept to a minimum while scheduling dental care, information from the hematology team regarding the patient's clinical status and most recent blood test results should be advised in order to conduct a thorough risk assessment. It is important to decide whether primary or secondary (hospital-based) care settings are the best for providing care. [9]

In the present case, a close association with pediatric hematologist, oncologist, cardiologist, anesthetist, pediatrician along with the pediatric dentist and his blood profile was maintained.

Guidelines according to Thalassaemia International Federation on dental management of beta thalassemia major patients were followed for an overall favorable outcome.[9]

CONCLUSION

The decision to extract in such a case, instead of any endodontic therapy was dictated by looking beyond the oral cavity thereby having a holistic approach towards the health of the child.

There is an absolute need to be a part of a dynamic multidisciplinary team when treating special health care needs children and if needed compromise on the dental treatment and shift from being dental centric to life centric.

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