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Ordeals in the Dental Management of a Child with Lesch Nyhan Syndrome: A Case Report

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ABSTRACT

Lesch Nyhan syndrome is a rare X linked recessive genetic disorder characterized by errors in purine metabolism, due to deficient activity of hypoxanthine guanine ribosyl transferase (HPRT). Lesch Nyhan syndrome is associated with three major signs; uric acid overproduction, neurologic dysfunction and cognitive, behavioral disturbances inducing self-mutilation. Hereby, we report a case of a 7 year old male child with Lesch Nyhan syndrome who reported to our department with ulcerations of the labial, buccal mucosa and tongue due to self-mutilation. The dental treatment conferred to alleviate the symptoms of self-mutilation are discussed in this case report.

Keywords: Acrylic splint, Lesch Nyhan syndrome, Oral ulcers, Self-mutilation.

INTRODUCTION

Lesch Nyhan syndrome is a rare congenital condition inherited as an X linked recessive trait that primarily affects males with females as carriers. [1] It is characterized by a severe deficiency of the enzyme hypoxanthine guanine phosphoribosyl transferase (HGPRT).[2]

Affected children typically have a normal prenatal and perinatal course. Hypotonia and developmental delay are evident from three months of age.[4] Pathognomonic general features include spasticity, hyperreflexia, and extensor plantar reflexes, motor disabilities and mental retardation. The most distinctive symptom is compulsive self-injury. The characteristic intra oral features manifest with the eruption of deciduous teeth, they begin to show self-mutilation, biting their oral/perioral tissue and fingers.[3]

The objective is to describe a case of Lesch Nyhan syndrome, the challenges faced, dental procedures performed and an overview of the available treatment modalities.

CASE REPORT

A 7 year old boy was referred to our Department of Pediatric and Preventive Dentistry by the neurologist for evaluation of occlusion and self-mutilation of the labial, buccal mucosa and tongue. Medical history demonstrated the patient to have Lesch Nyhan syndrome grade 4 confirmed by biochemical and chromosomal analysis. Severe global developmental delay with motor, language, social and intellectual deficits were present. Patient is on allopurinol to prevent accumulation of uric acid. Family pedigree revealed the patient to be an only child born to non-consanguineous parents with no known history of

Lesch Nyhan syndrome in the family. Parents reported first signs of self-mutilation at one and a half years of age. General examination revealed a scooped out raw wound on the right middle finger and partially healed wound on the left acromion joint. (Figure 1)

Intra oral examination showed multiple bleeding ulcers covered by slough on the right and left buccal mucosa, lower labial mucosa, hard palate, lateral borders of the tongue (Figure 2). Parents revealed a history of consulting a dental hospital when the patient was 5 years old, during which time a pair of intra oral resin mouth guards were given. The patient was able to remove both mouth guards within a few seconds of placement and did not comply to wear the mouth guards thereafter. This was taken into consideration during our treatment planning and the decision for a fixed appliance was undertaken.

Irreversible hydrocolloid impressions of both arches were taken, casts poured. A hard acrylic splint was fabricated, cemented with type I Glass ionomer cement in relation to 11, 21 to provide inter occlusal clearance in the posterior region and prevent cheek, lip biting. Keeping in mind the age of the patient and eruption sequence, maxillary deciduous lateral incisors were excluded from coverage by the splint. Composite bite blocks were placed on 73,74,75,83,84 and 85 with contact in the functional cusp region and clearance in the non functional cusps in order to stabilize occlusion and provide a tripod contact. (Figure 3) Pit and fissure sealants were done on 36, 46 with type IV Glass ionomer cement.

As an aid to prevent self-mutilation of the fingers, a pair of hand-finger control mitts protection gloves with loop closure were advised.

Follow up examination in one week revealed healing of oral ulcers and decrease in self-mutilation of the oral cavity. Prohibition of intercuspation of maxillary and mandibular posterior teeth prevented the entrapment of buccal mucosa and tongue.

Parents were advised to maintain oral hygiene and recall visits every month. One month follow up showed a complete healing of the ulcers and remission of oral self- mutilating behaviour with the parents, highly satisfied with the treatment. The splint and composite bite blocks were removed in three months to prevent possible occurrence of Temporomandibular disorders due to the open bite caused by the splint. The parents were advised about recementation if the habits recur. Six months follow up revealed mild episodes of cheek and lip injuries which were managed by placement of composite bite blocks on the mandibular posterior teeth and palliative measures.

DISCUSSION

Self-mutilating behaviour is seen in a number of conditions like Tourette syndrome, Manchausen syndrome, Lesch Nyhan syndrome, Cornelia de Lange syndrome, Rett syndrome.

Lesch Nyhan syndrome is a HPRT1 disorder, characterised by over production of uric acid resulting in a myriad of neurological and behavioural problems. Three phenotypes have been identified in the spectrum of HPRT 1 disorders, ranging from mild to severe. Lesch Nyhan syndrome being at the more severe end can again be categorized into 4 variants based on the degree of compromise as illustrated in Table 1.[3] The present case report is of a child diagnosed with Lesch Nyhan syndrome, grade 4.

The various treatment modalities to manage oral self-mutilation in Lesch Nyhan syndrome include conservative and invasive procedures, namely, removable soft mouth guards, bite blocks, a combination of extraoral and intraoral orthodontic apparatus that covers the chin and is held in place with a helmet on the neck-strap, shields that protect the tongue and lips from direct injury, labial bumpers welded onto orthodontic bands or stainless-steel crowns, total extraction.[4] Dabrowski and Medicine reported a case wherein Injection of Botulinum toxin A in the masseter muscle prevented presynaptic release of acetylcholine, causing motor plaque dysfunction and muscle weakness with a consequent reduction in self-mutilated oral lesions.[5] Macpherson et al. described a child with Lesch Nyhan syndrome treated with orthognathic surgery and an open bite was created, limiting the patient's ability to chew.[6]

In terms of total extraction, it should be kept in mind that once extracted it cannot be undone. Goodman; Gonzalez et al have reported total extraction to be successful, while Livia Gisbert de la Cuadra et al reported total extraction to have limitations in preventing self-mutilation. [7,8]

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In our patient a conservative approach was followed owing to the age of the patient, parents' wishes, and dentition status. Some of the challenges faced during treatment planning were; removable soft mouth guards, the most common conservative treatment modality had already proved to be inconsequential, patient's dentition was in the first transition period, highly aggressive behaviour of the child, parents' unwilling for sedation due to which all procedures were carried out with physical restraints.

The characteristic behavioural phenotype in Lesch Nyhan Syndrome is biting of fingers which was in accordance with our patient. Treatment modalities suggested in literature apart from protection mitts to control extra oral self-mutilation are long sleeved shirts and music therapy.[9]

CONCLUSION

Every child with Lesch Nyhan syndrome is unique and one specific treatment plan cannot be advocated for all patients. Allowances must be made for parents' views and inclinations. Despite the challenges involved, conservative treatment approaches should be exhausted before considering total extraction. More literature in this regard will enable pediatric dentists to confer a holistic treatment for such children thus vastly improving their quality of life.

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TABLES/FIGURES

Grade 1	No neurological manifestations, independent and autonomous, dystonia while exercising, slight attention deficit disorder and obsessive behaviour.
Grade 2	Mild neurological symptoms, independent walking, macrocytic anemia, mild intellectual disability
Grade 3	Acute neurological symptoms, acute generalized dystonia, no self-mutilating behaviour
Grade 4	"Classic Lesch-Nyhan Syndrome", acute generalized dystonia, ballism, self-aggressive behaviour, cognitive impairment, megaloblastic anemia

Table 1: Variants of Lesch Nyhan syndrome[4]

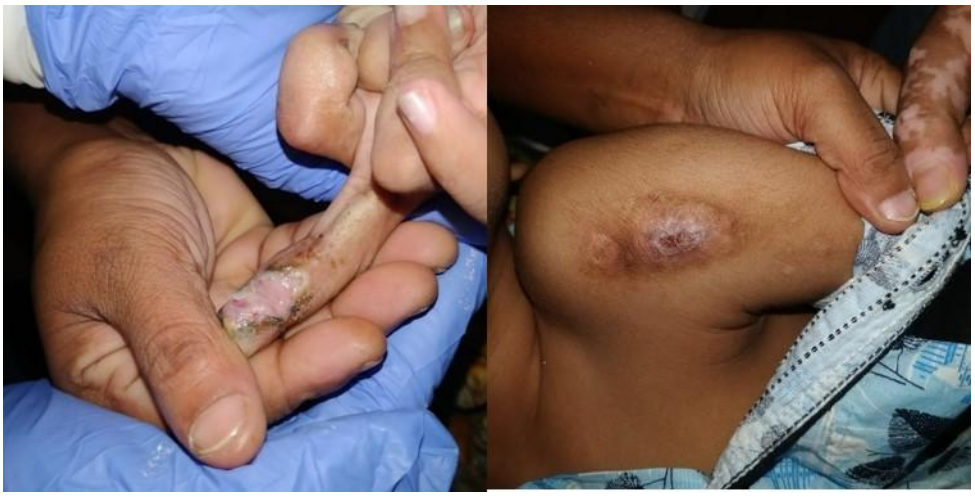

Figure 1: General examination

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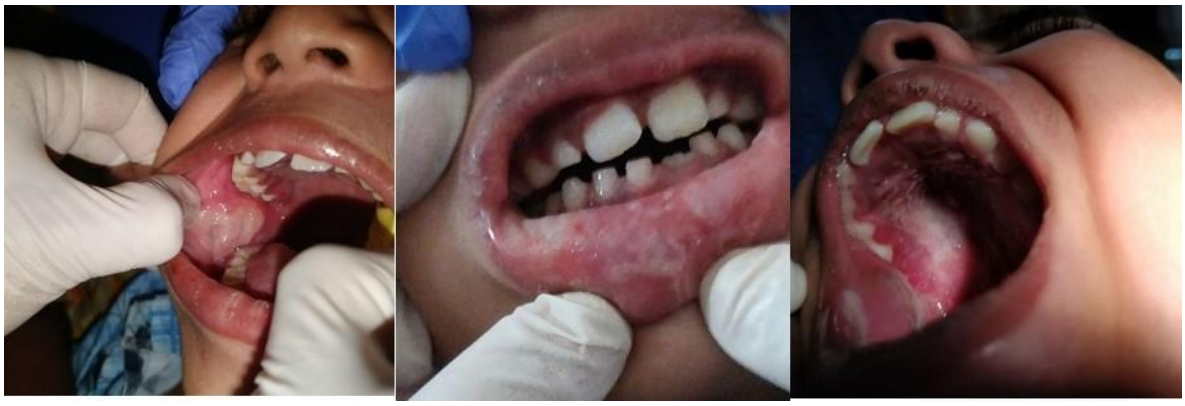

Figure 2: Intra oral features

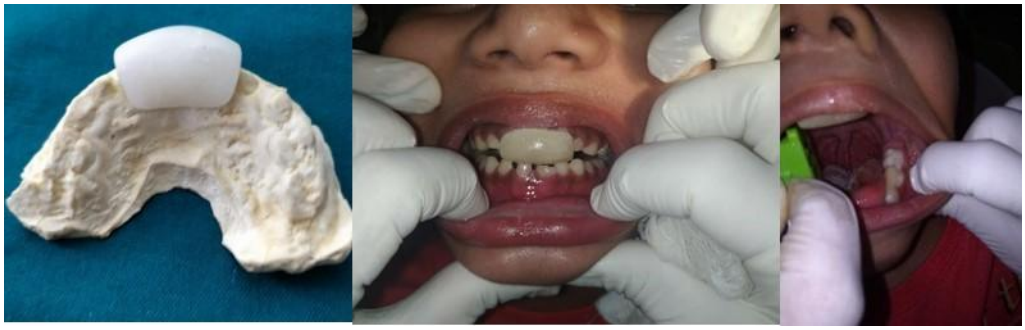

Figure 3: Acrylic Splint and Composite bite block

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