Surgical Management of Self-Mutilation Due to Lesch-Nyhan Syndrome in a Pediatric Patient

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BACKGROUND

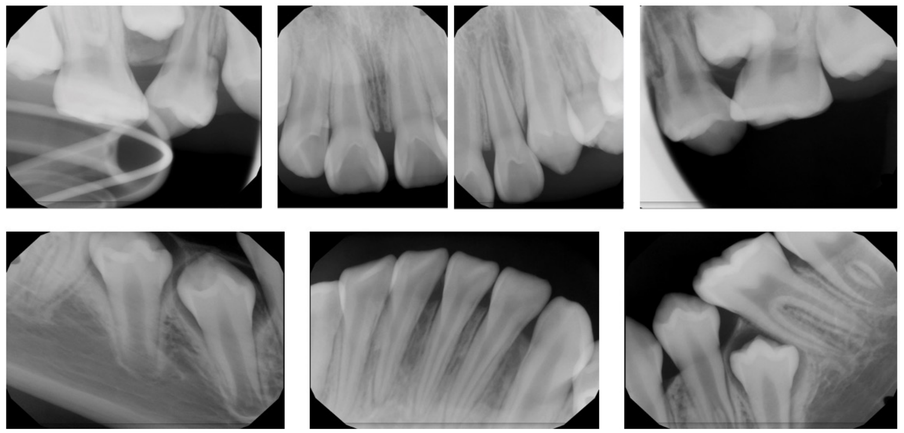
Lesch-Nyhan Syndrome (LNS) is an X-linked recessive disorder that disrupts the metabolism of purines caused by a mutation in Xq26.2-q26.3, which leads to an increased buildup in uric acid due to a deficiency in the HGPRT enzyme[1]. Genetic testing and pedigree analysis along with self-mutilative behavior aids in confirming the diagnosis of LNS. Patients often exhibit neurological and neurocognitive deficiencies, with a lack of neuromotor control[2]. Gout and kidney issues such as nephrolithiasis are also reported. The self-mutilating behavior seen in LNS often involves the fingers as well as the lips and tongue. Repetitive trauma to the patient's lip can lead to the formation of a fibroma, and in some instances, tissue necrosis leading to loss of function or loss of structure.

CLINICAL PRESENTATION

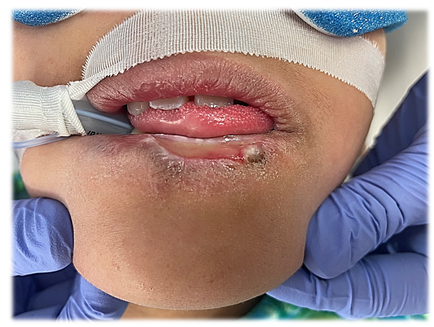
A 10 year, 4-month-old male presented to Riley Children's Hospital Dental Clinic as a new patient for a comprehensive examination after his father noticed extensive trauma to the patient's lower lip. His medical history includes Lesch-Nyhan Syndrome, quadriplegia, nephrolithiasis, developmental/learning delay, g-tube fed, cerebral palsy, and muscle spasms. He takes allopurinol, baclofen, Sinemet, and SAM-e (S-adenosylmethionine). He has no known drug allergies. The patient had previously undergone full-mouth extraction of the primary dentition due to self-mutilating behavior. The patient presents with permanent dentition, but teeth #20, 28, and 29 are unerupted due to impaction.

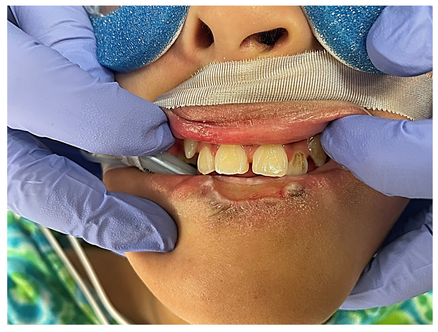
The patient had a history of continuous lower lip biting, which now presented as a large, ulcerative lesion with irregular borders and a white fibrin cap in certain areas. The lesion was scarred and very taught, making manipulation of the lower lip challenging. The patient's father stated the patient's lower lip appeared to continuously bother him due to tightness and ulcerations. A treatment plan was proposed to address the self-mutilation using surgical intervention.

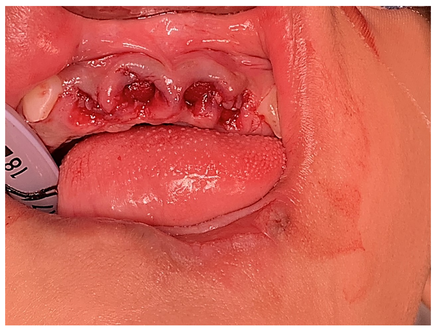
RADIOGRAPHS

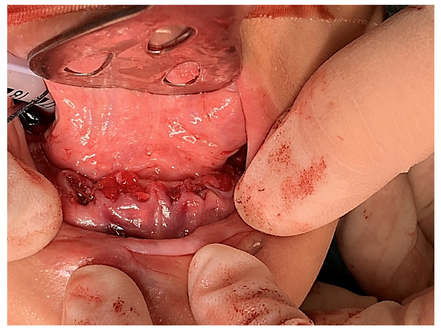


CLINICAL PHOTOGRAPHS



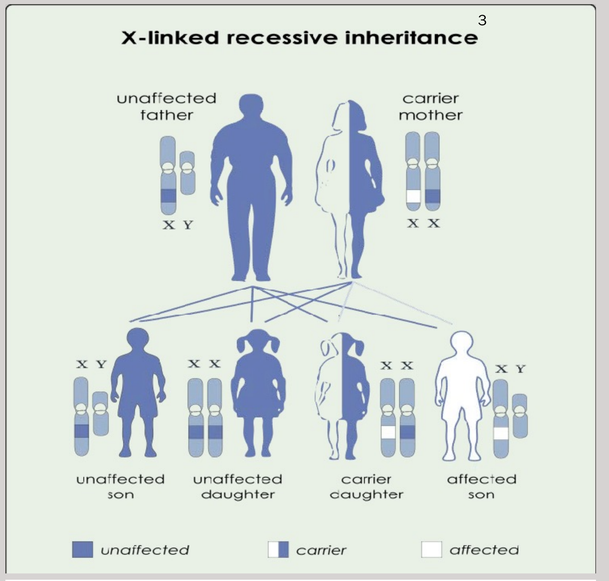






TREATMENT/MANAGEMENT

The patient was brought to Riley Children's Hospital Day Surgery where comprehensive dental treatment was completed under general anesthesia. An oral evaluation was completed, radiographs were taken, and a treatment plan was composed and completed. Teeth #4, 7, 8, 9, 10, 13, 21, 22, 23, 24, 25, 26, 27 were extracted with no complications followed by a prophylaxis and application of 5% NaF varnish. The patient was provided a recall appointment in 6 months in the outpatient clinic for a periodic oral evaluation and to assess the condition and healing of the lower lip. Studies have stated mouth guards can be attempted prior to surgical intervention along with consistent follow-up visits[4]. The outlook for proper tissue healing is good once the patient is treated successfully via mouth guard or surgical intervention.



REFERENCES