Gingival Fibromatosis in a Pediatric Patient with Zimmermann-Laband Syndrome

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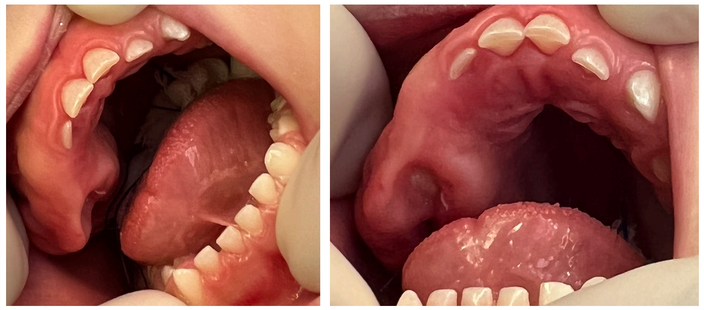
ABSTRACT

A 5-year-old female presented to the dental clinic at Riley Hospital for Children for initial examination, routine recall examinations and any necessary dental treatment. Her health history includes Zimmermann-Laband syndrome and history of seizures. The patient is non-verbal with limited use of her extremities and in a wheelchair. She is taking oxcarbazepine for seizures and is allergic to amoxicillin and clindamycin. The patient had an initial exam in October 2019 and was monitored with 6-month recalls until April 2021. In April 2021, the patient's mother reported difficulty eating with probable pain and only eating a soft diet due to gingival overgrowth in the upper right. The patient's mother was provided treatment options and agreed to gingivectomy treatment with biopsy under general anesthesia. Treatment was completed in the operating room at Riley Hospital for Children in December 2021. This report will include discussion of Zimmermann-Laband syndrome and treatment for hereditary gingival fibromatosis including differential diagnoses, radiographic and histological findings.

BACKGROUND

Zimmermann-Laband syndrome is a rare genetic disorder. Research suggests that it is an autosomal dominant trait, but some evidence shows autosomal recessive inheritance. The syndrome is characterized by craniofacial and hands and feet abnormalities. Facial abnormalities include narrow facial appearance with increased size of tongue, lips, nose and/or ears. Children with Zimmermann-Laband syndrome also commonly present with overgrowth of gingival tissue. This gingival enlargement may completely cover the teeth which can cause malocclusion, chewing problems, excessive salivation and/or difficulty swallowing. Treatment includes gingivectomy to uncover the dentition and likely future recurrence due to Zimmermann-Laband syndrome.[1]

CLINICAL INTERPRETATION



Initially, a clinical examination was completed, and photographs were taken. Examination reveals firm, generalized enlargement of the right posterior gingiva, extending bucal-lingually from the distal of #A to the distal of #D. The enlargement was classified as Grade 3, covering ¾ or greater of the crowns of the right posterior teeth. The initial clinical impression was gingival hyperplasia.

DIFFERENTIAL DIAGNOSIS

The American Academy of Oral Medicine (AAOM) classifies generalized gingival enlargement into four categories: Inflammatory-induced, medication-induced, systemic disease-induced, and hereditary or idiopathic gingival fibromatosis.[4]

Inflammatory-induced gingival enlargement occurs from plaque, poor oral hygiene, and/or poor restorations or orthodontic appliances.

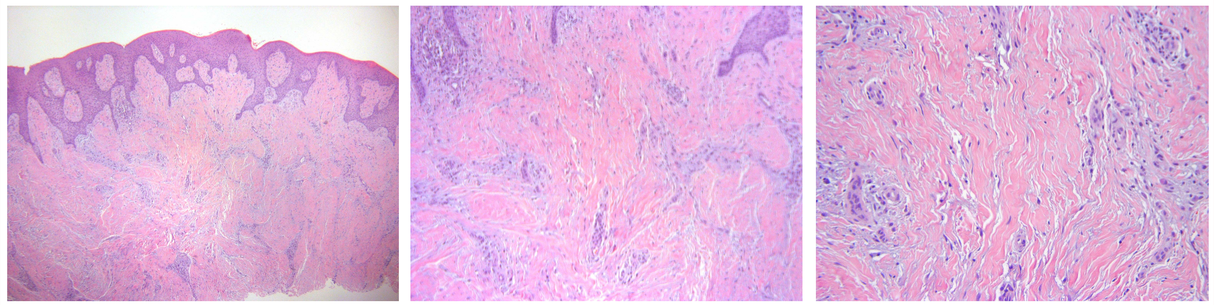
Medication-induced gingival enlargement occurs with multiple drugs. These include anticonvulsants such as phenytoin (50% prevalence), calcium channel blockers such as nifedipine (25% prevalence) and immunosuppressants such as cyclosporine (25% prevalence). The mechanism of action for medication-induced gingival enlargement is an increase in gingival fibroblasts causing a decrease in collagen degradation leading to an increase in connective tissue matrix.[2,3]

Systemic disease-induced gingival enlargement can occur due to leukemia. Gingivitis occurs secondary to neutropenia and further gingival enlargement occurs due to leukemic infiltrates. Other signs of this enlargement include spontaneous bleeding, petechiae, ecchymosis, ulcerations and mobility of teeth.[3]

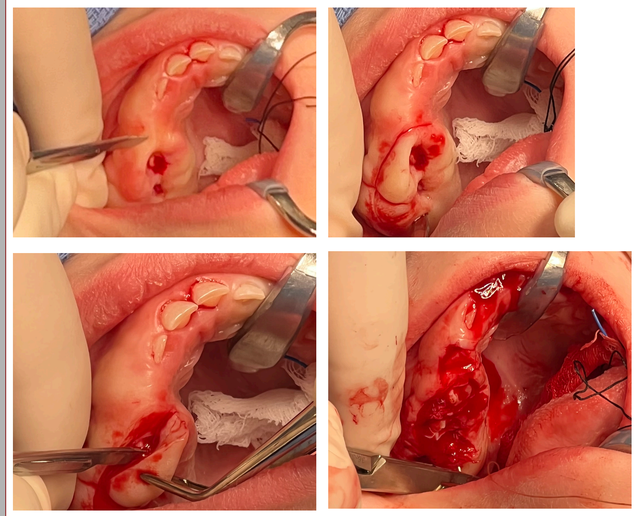
BIOPSY AND PATHOLOGY REPORT

A pathology report and photographs were provided by Dr. Paul Edwards, MS, DDS from the Oral Pathology Group at Indiana University School of Dentistry. The original clinical impression was gingival hyperplasia. The pathology report stated, "Histologic examination reveals hyperplastic oral mucosa surfaced by focally hyperplastic stratified squamous epithelium overlying dense fibrocollagenous tissue, with interspersed focally myxoid areas, occasional dystrophic calcifications surrounded by multinucleated giant cells and epithelial islands. Stellate-shape fibroblasts and focal multinucleated fibroblasts are also identified." The final diagnosis was right maxillary gingiva, gingival fibromatosis.

According to Ragaei and Abdul Moneim (2017), the normal histopathologic features of hereditary gingival fibromatosis include epithelial hyperplasia, elongated rete ridges extending into the underlying connective tissue, and excess collagen in the connective tissue with activated fibroblasts present. Small osseous calcifications and abundant neurovascular bundles may also be present.



TREATMENT



Following clinical examination and consultation with attending faculty, Dr. Juan Yepes, it was determined the patient would best be treated in the operating room under general anesthesia. A gingivectomy was completed uncovering the crowns of the right maxillary primary posterior teeth. All gingival specimens were submitted for histological examination in 10% neutral buffered formalin to the Oral Pathology Group at Indiana University School of Dentistry. Hemostasis was key prior to extubation due to the extensive area of resection. Positive pressure and multiple 3-0 chromic gut sutures were placed to aid with hemostasis.

Post-operative instructions to the patient's parent were vital. Post-operative pain and bleeding may occur following treatment. Continued follow-up care is important. A 2-week re-evaluation is advised to evaluate healing. Continued 6-month recall examinations are important to evaluate the gingival enlargement for recurrence.

CONCLUSIONS

Hereditary gingival fibromatosis is a common occurrence in patients with Zimmermann-Laband syndrome as well as other inherited syndromes including Rutherfurd, Cross, Cowden, Costello and Ramon syndrome.[4]

These syndromes can be associated with other clinical manifestations such as hypertrichosis, epilepsy, mental deficiency, growth retardation, hypopigmentation, optic and auditory defects, flexible joints, and heart and craniofacial abnormalities.[4]

According to the literature, the treatment completed is in line with the recommended treatment. Hereditary gingival fibromatosis is indicated for surgical excision and gingival recontouring with biopsy.[3]

Unfortunately, due to this patient's gingival enlargement being diagnosed as hereditary gingival fibromatosis, there is likely recurrence of the enlargement.[3]

REFERENCES

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