Case Report on Dental Management of a Pediatric Patient with Overgrowth Syndrome

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INTRODUCTION

Overgrowth syndromes are a group of genetic disorders that cause excessive generalized or localized growth before and after birth[1]. Various overgrowth syndromes are associated with hormone imbalances, life-threatening hypoglycemia, seizures, developmental delay, and an increased susceptibility to malignancy[2]. There is very little literature that describes dental manifestations seen in patients with any of the overgrowth syndromes.
This is a case report of interdisciplinary management of a pediatric patient with overgrowth syndrome who presented to the pediatric dental clinic at One Brooklyn Health Brookdale Hospital Medical Center.

HISTORY

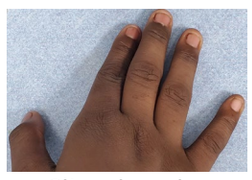
An 11-year-old male presented to the pediatric dental clinic (referred by the Emergency Department) for pain and swelling near the lower left posterior teeth. Patient had previously been seen in the dental clinic but did not follow up for continuing care.
Medical History:

- Overgrowth syndrome (differential diagnosis based on history and clinical presentation; genetic evaluation and genetic testing pending)
- Weight: 149 lbs; Height: 5’ 6’’
- Precocious puberty
- Autism spectrum disorder
- H/O Ventricular Hypertrophy
- Mild Persistent Asthma

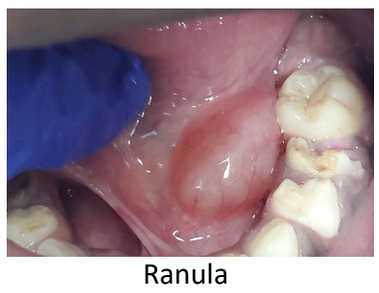
CLINICAL EXAM

Almond shaped slanting palpebral fissures
Epicanthal folds
Thick eyebrows

Narrow frontal width
High forehead



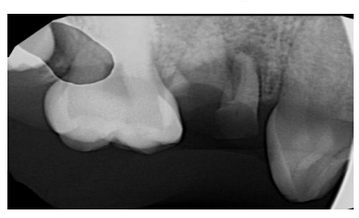
Clinodactyly



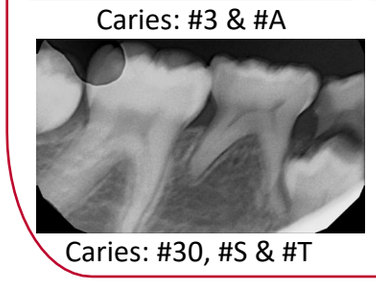
Ranula

RADIOGRAPHS

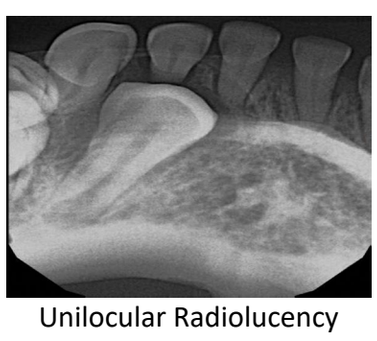
Oligodontia (#4,5,7,10,13,16,17,20,21,22,23,24,25,26,29,32)



Caries: #3 & #A



Caries: #30, #S & #T



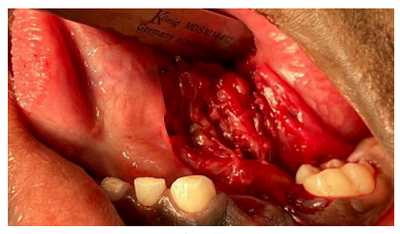
Unilocular Radiolucency around #27

Caries: #14 & #J

Caries: #19, #K & #L

PERI OPERATIVE AND POST OPERATIVE PHOTOS

Pre-op Ranula
(On day of surgery)



Post-Ranula Excision
(Prior to suture placement)

Post-Op Ranula
(On day 12 post-op)

Post-op Extraction Socket
(Area of cyst - On day 12 post-op)

CASE MANAGEMENT

1st visit (emergency visit)

Pain management, patient discharged with oral antibiotics prescribed in ED

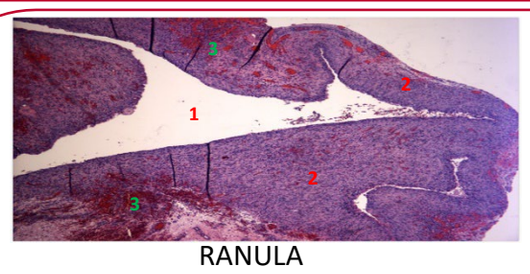
2nd visit (comprehensive exam)

- Oral surgery consult for evaluation of ranula
- Patient planned for comprehensive oral care under general anesthesia due to limited patient cooperation and benefit from multidisciplinary treatment in the operating room.
- Prosthetic consult and tentative treatment plan for missing teeth:
    1. Provisional restoration with maxillary and mandibular RPDs until #27 and #28 are erupted
    2. Evaluate for implants or fixed partial dentures in the future
- Medical consult (pulmonology and cardiology): patient cleared for treatment under general anesthesia, no antibiotic prophylaxis recommended.

3rd visit (Comprehensive treatment under general anesthesia with Oral Surgery

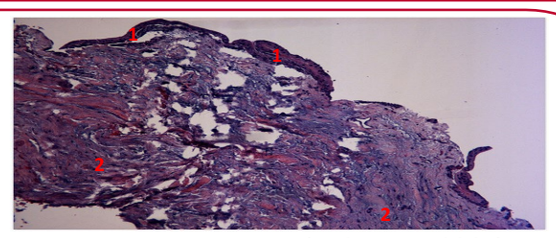
- Composite restorations: #3, 14, 19, 30
- Stainless steel crowns: #J and #T
- Extraction: #A, K, L, P, Q, S ; and excision of cystic lesion around #27
- Excision of ranula and the entire left sublingual gland

PATHOLOGY



RANULA

1. Ranula without mucin
2. Compressed Granulation Tissue containing Macrophages
3. Extravasated RBCs due to surgical incision



DENTOGEROUS CYST (right mandible)

1. Epithelial lining
2. Fibrous Connective Tissue Wall

COMMON OVERGROWTH SYNDROMES

Syndrome	Characteristics
DNMT3A-related[2,6]	Macrocephaly, round face, thick eyebrows, prominent maxillary incisors, intellectual disability, autism spectrum disorder, heart defects, joint hyperlaxity
Sotos[1,2,4,5]	Seizures, macrocephaly, tall and narrow skull, downward slanting eyes, high forehead, pointed chin, cognitive impairment, joint hyperlaxity, supernumerary teeth
Beckwith–Wiedemann spectrum (BWS)[1,2,3]	Macroglossia, macrosomia, life-threatening hypoglycemia, cardiac anomalies, ear creases/pits
Simpson–Golabi–Behmel[1,2]	Similar to BWS with facial features becoming prominent with age, extra nipples
Perlman[1,2,7,8]	Macrosomia, macrocephaly, hypotonia, nephromegaly, high risk for Wilms tumor
PTEN hamartoma tumor syndromes[2,9]	Multiple hamartomas. Increased risk of tumors in Cowden syndrome – thyroid, breast, kidney, endometrium, colorectal and skin
PIK3CA related overgrowth spectrum[1,2,10]	Lipomas, vascular malformations, overgrowth of extremities, scoliosis, megalencephaly
Proteus syndrome[2,11]	Progressive overgrowth of feet and/or hands, connective tissue nevi having mosaic pattern

RANULA

Dome-shaped, fluctuant swelling with blush hue in the floor of the mouth, caused by rupture of sub-lingual gland ducts[12]. Treatment includes sclerotherapy, aspiration, marsupialization, sole excision of ranula or excision of ranula along with sublingual gland[13,14,15,16].

DENTIGEROUS CYST

Encloses the crown of an unerupted tooth, attached at the CEJ. Appears as a well-defined unilocular radiolucency surrounded by a sclerotic border in radiographs. Treatment includes marsupialization, enucleation or curettage[12].

CASE DISCUSSION

- Patient presents with autism spectrum disorder, round face, thick eyebrows and heart defects which are commonly seen in DNMT3A-related overgrowth syndrome. Genetic testing needs to be done for a definitive diagnosis.
- Excessive growth of tissues and organs is seen in overgrowth syndromes. Cardiology and pulmonology consult is needed prior to dental treatment, especially under general anesthesia.
- Due to patient’s behavior, severe caries pattern, medical condition, and pathology, more definitive and predictable treatment was done to minimize the risk for recurrent caries, infection, and other complications.
- Association among oligodontia, cystic lesions, and overgrowth syndromes has not been reported in the literature. The clinical presentations in this patient may be a manifestation of the underlying genetic abnormality.
- It is important to retain as many teeth as possible to preserve bone for future prosthetic restorations.
- Preventive care includes 3-month recalls, fluoride varnish applications, using Prevident-5000 toothpaste. Emphasized the importance of oral health care to the parent.

REFERENCES

Presented at the 98th Annual Session of the Greater New York Dental Meeting in 2022