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Ehlers Danlos Syndrome with Multifarious Oral Manifestations: A Case Report

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Abstract: ***Ehlers - Danlos syndrome (EDS) denotes a group of heritable disorders of connective tissue consisting of mutations in the genes involved in the production of collagen. EDS being a disorder affecting collagen, a prime component of tooth and supporting structures, results in various dental anomalies. A 14 year old female patient with a diagnosis of autosomal dominant classical Ehlers Danlos Syndrome, reported to our Department. This case report showing various oral manifestations present in this patient and the treatment we rendered.***

Keywords: Classic Ehlers Danlos Syndrome, Oral Manifestations, Midroot Fracture, Dental Management, Case report

1. Introduction

Ehlers - Danlos syndrome (EDS) describes a group of heritable disorders of connective tissue consisting of mutations in the genes involved in the production of collagen. [1] The syndrome is estimated to affect about 1 in 5, 000 to 100000 live births in different communities. [2]

The molecular basis of classic EDS is a deficiency of type V collagen, a quantitatively minor fibrillar collagen that is widely distributed in tissues such as skin, bone, tendon, cornea, placenta and foetal membranes. It comprises three different α - chains encoded by the COL5A1, COL5A2 and COL5A3 genes, respectively. [3] EDS being a disorder affecting collagen which is a prime component of tooth and supporting structures results in various dental deformities and oral manifestations.

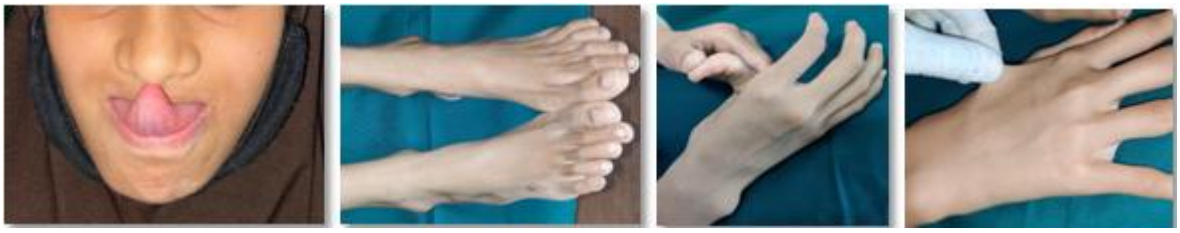
2. Case Report

A 14 year old female patient, who was a previously diagnosed case of classic Ehlers Danlos Syndrome (cEDS), reported to our Department of pediatric and Preventive Dentistry. Patient presented with pain and sensitivity in the lower front teeth region since 3 months. Child was born out of consanguineous marriage with an insignificant prenatal and natal history. She presented loose joints and delayed wound healing since early childhood along with prominent down slanting palpebral fissures. She was undergoing physiotherapy related to frequent dislocation of left and right shoulder joint.

Patient gave history of orthodontic treatment followed by a fixed lingual splint in relation to the lower anteriors, which were mobile after completion of orthodontic treatment, along with multiple restorations which were done 3 years back. Extra oral examination (Figure 1) revealed positive gorlin sign, prominent down slanting palpebral fissures, hyper extensible and smooth velvety skin, hypertrophic scar on leg and joint hypermobility. Intra oral examination (Figure 2) revealed generalized enamel hypoplasia, dental caries in relation to 16, 33, 32, 31, 36, 42, 43, 46, 21, 22, 23, 26, 27, Chronic marginal gingivitis, deep pits and fissures, posterior cross bite in the right permanent second molar region, fixed lingual splint in relation to mandibular anterior region from 34 to 44.

Radiographic investigations with IOPAR, OPG and CBCT (Figure 3) revealed horizontal midroot fracture in relation to 41 and morphological variations of root in relation to 31, 32, 33, 42, 43, along with significant bone loss in the mandibular anterior region and pulp stones in relation to 37 and 47.

An appropriate restorative/endodontic treatment plan was designed and presented to the parents and an informed consent was obtained. Oral prophylaxis followed by Composite restorations of carious teeth and pit and fissure sealant application of partially erupted second molars along with topical fluoride application were done (Figure 4). Endodontic treatment in relation to 41 was carried out.



(a) (b) (c) (d)

Figure 1: a) Gorlin sign. **b&c)** Hypermobile joints. **d)** Smooth and velvety hyperextensible skin

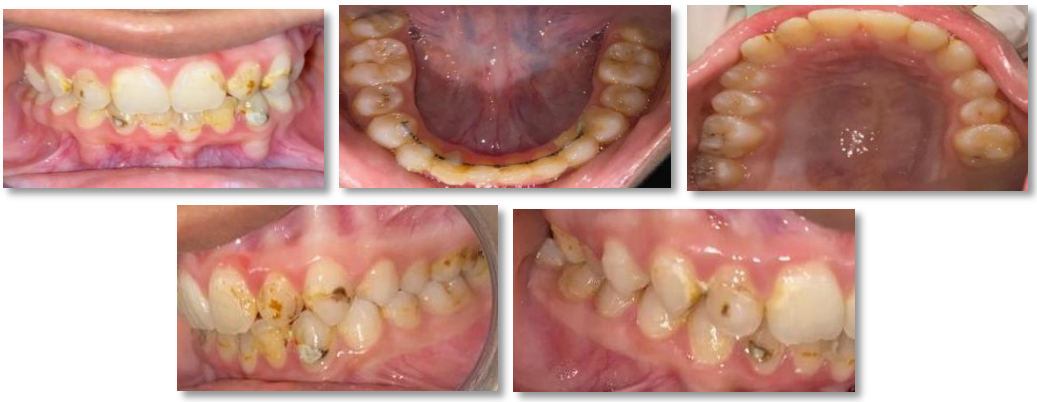


Figure 2: Pre - operative images

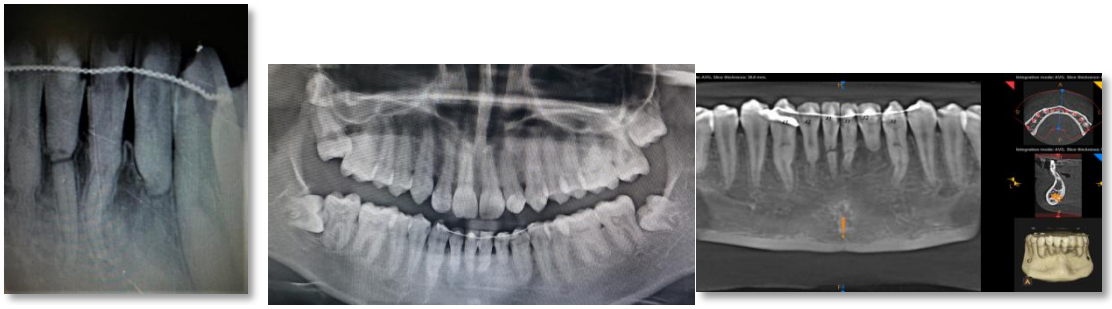


Figure 3: Showing IOPAR, OPG and CBCT images

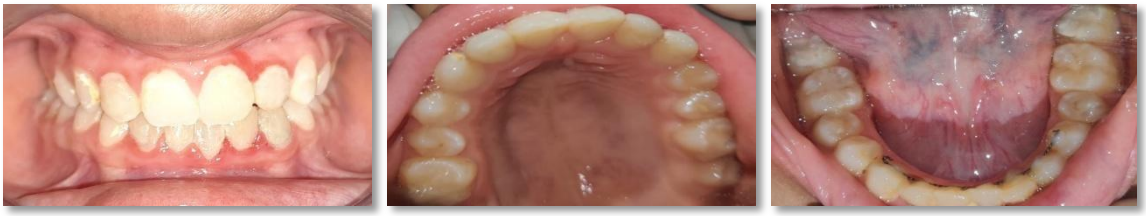


Figure 4: Post - operative images

3. Discussion

The oral manifestations of EDS include fragile mucous membrane, dentinal aberrations like pulp stones, short and deformed roots, high incidence of caries in the deciduous teeth, Spontaneous fractures of teeth, Early onset of generalized periodontitis, high - arched palates and supple tongue. [4] Gorlin and Pindborg reported 50% of the patients with EDS had the ability to touch their nose with their tongue (Gorlin sign), versus the 8% to 10% of the normal population. [5] Major diagnostic criteria for classical EDS includes skin hyper extensibility, wide atrophic scars, and significant joint hypermobility. [6] Similar clinical signs were observed in the presented case. TMJ hypermobility and high chances of dislocation should be considered while doing dental treatments [7] and short duration of dental appointments are preferred for such patients.

Pillsbury showed that fragility of skin and blood vessels could lead to scar and hematoma formation following any traumatic event in EDS patients. [8] To avoid complications, supragingival scaling was performed in this patient instead of subgingival scaling. One of the most important findings in this patient was a horizontal midroot fracture in relation to 41. Since the patient did not give any history of trauma, the fracture could have occurred because of the nature of the disease itself along with the force applied during her previous orthodontic therapy.

Andreasen and Hj¨orting - Hansen described four types of healing sequelae for midroot fracture:
(1) healing with calcified tissue, (2) healing with interproximal connective tissue, (3) healing with interproximal bone and connective tissue, and (4)

interproximal inflammatory tissue without healing. [9] In the present case biodentine was used for endodontic therapy to induce apposition of reactionary dentine by odontoblast stimulation and reparative dentin by cell differentiation. [10] A long term follow - up is necessary to know the improvement in fracture healing and the patient recalled every 3 months for evaluation.

4. Conclusion

Pediatric dentistry practitioners must know all relevant oral and general manifestations of Ehlers Danlos syndromes for early detection and to provide adequate oral management. There are instances where we are the first line health care providers these children come across for their dental problems. Equipping ourselves with the knowledge of such syndromes will lead to early referral and diagnosis and it is crucial for the positive health of the patient. Therefore, the pediatric dentist should be part of a multidisciplinary health team in order to perform a precise and opportune diagnosis.

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