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Ectrodactyly Ectodermal Dysplasia Cleft Patient Management in Pediatric Dental Clinic - A Case Report

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Abstract

Ectrodactyly Ectodermal Dysplasia Cleft syndrome is a rare disorder with the triad of split-hand or leg with the missing of the central digits, defect in hair, skin, dental tissue, salivary glands, and Cleft lip and or palate or only Cleft Palate alone. The incidence of this syndrome is 1.5 in 100 million populations. Patients with EEC syndrome have poor oral hygiene because of compromised dexterity of the upper limb. This is a case report describing the conservative management of the carious tooth in the EEC syndromic patient for the preservation of the alveolar ridge for future prosthetic rehabilitation.

Keywords: Ectrodactyly Ectodermal dysplasia, Ectodermal dysplasia, Cleft Lip and Palate.

Introduction

Ectrodactyly ectodermal dysplasia Cleft (EEC) was first described by Eckoldt and Martens in 1804 and Rudiger et al. in 1970 coined the term EEC syndrome [1,2]. Ectrodactyly ectodermal dysplasia Cleft syndrome is an autosomal disorder with the triad of ectrodactyly / split hand or split foot malformation, ectodermal dysplasia and Cleft lip and /or palate. Ectodermal dysplasia (ED) comprises defects in the derivatives of embryonic ectoderm and present as hypodontia or oligodontia,

dystrophic nails, dry skin, or sparse hair [3]. Ectodermal dysplasia results in a defect in the skin, hair, nail, salivary glands, oral soft and hard tissues. The incidence of this syndrome is 1.5 in 100 million populations [4]. Other features associated with this syndrome are hearing loss, lacrimal duct defect, genitourinary defect, delayed developmental milestones, malignant lymphoma, and occasionally mental retardation [5].

The teeth in this syndrome might also be hypo-plastic in nature and more prone to post-eruptive breakdown and dental caries. Here is a case report of a patient with EEC syndrome who reported to the pediatric dental clinic and its management before the prosthetic rehabilitation has been explained.

Case report

An 8 years old Female child presented to the Department of Pedodontics and Preventive Dentistry, with the chief complaint of multiple missing teeth. They also complained of limb anomalies. The Family history was irrelevant. The patient was the elder daughter followed by her was a boy baby who was normal. There was a past medical history of cleft lip and palate operated at 6 months and 3 years. Dental history of extraction of 36 was done in a private clinic. Physical examination revealed lightly pigmented hair, dry skin [Figure 1]. The upper limb examination revealed a split hand of both the right and left limb with the middle finger missing while in the lower limb it was syndactyly with the fusion of the toe fingers [Finger 2]. In the left foot, there was fusion of second and first toe fingers and in the right foot, second and third toe fingers were fused [Figure 3]. Intraoral examination revealed decreased saliva secretion, oral mucositis, oligodontia with root stump of 54, 53, and 63, Carious 16, 26, 46, and hypo-plastic 11,21 [Figure]. The carious 46 was appreciated with grade II mobility and gingival swelling. The alveolar ridge in the lower arch was knife thin with inadequate height and width and classified as Class V according to Atwood classification. Panoramic radiograph revealed impacted 14, 24, 34, 44, 47 [Figure]. Treatment planning was done as preventive approach managing root stumps, carious and hypo-plastic teeth.

Treatment plan

The root stumps 54, 53, and 63 were extracted under Local anaesthesia with a vasoconstrictor (Lidocaine, 1:80000 adrenaline). Modified Hall technique was used to restore the 16 and 26. The carious lesion in 16 and 26 was excavated with the high-speed air rotor with water spray then Glass ionomer material (Ketac Molar, 3M ESPE) was packed. The stainless-steel crown (3M ESPE) of the appropriate size was cemented on 16, 26 with luting GIC (GC Gold Label Type 1) [Figure].

IOPA of the lower right molar (46) revealed periapical radiolucency and was diagnosed as periapical abscess [Figure]. Under local anaesthesia (Lidocaine, 1:80000 adrenaline), the access opening was done with diamond round bur and Biomechanical preparation done using hand files till 40 size k-files. The triple antibiotic paste was mixed with 2% chlorhexidine and placed in the canals of 46. After 2 weeks, 46 was obturated using Gutta Percha taper 2 %. The permanent restoration was packed in the access cavity followed by which the Stainless-steel crown was placed on 46. Hypo-plastic 11,21 was re-contoured with the composite resin (Ivoclar Vivadent Tetric Ceram N Composite) [Figure].

Discussion

EEC syndrome can be classified into two types based on the cleft in the oral cavity as cleft lip with or without cleft palate and cleft palate alone [6]. In this case, the patient has both cleft lip and palate. This disorder occurs due to mutations in a gene encoding p63 [7]. Patients with EEC syndrome are more vulnerable for poor oral health

and dental caries because of reduced salivary secretion and syndactyly their dexterity in maintaining proper oral health is inhibited [8]. These patients may also be malnourished due to lack of teeth which directly affects the mastication process. In this case, the child had a maxillary deficiency and unerupted 34, 44, 47 so the prosthetic rehabilitation was planned after the eruption of these teeth. In the anterior region of the lower arch, the alveolar ridge was knife thin with inadequate height and width so, after the eruption of premolars, and second right molar implant-supported denture was recommended. The dental mani festations of this syndrome persist throughout life, dentists should work diligently for the appropriate rehabilitation of the patient. The teeth present in the oral cavity are mostly conserved in order to preserve the alveolar bone for support and retention of the prostheses. In this patient, it was planned for the implant-supported prostheses along with many surgeries to improve the vestibular depth and to increase the height of the alveolar ridge which is planned after the eruption of lower premolars and right lower permanent molar.

Conclusion

It is always vital to follow conservative management in these syndromic patients to preserve the alveolar ridge. One of the duties of the dentist is to have a clear awareness of the treatment procedures and the complications encountered.

This case report is described in order to provide knowledge regarding the EEC syndrome and to incorporate the idea of preserving the alveolar ridge by retaining the erupted teeth.

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Legend Figures

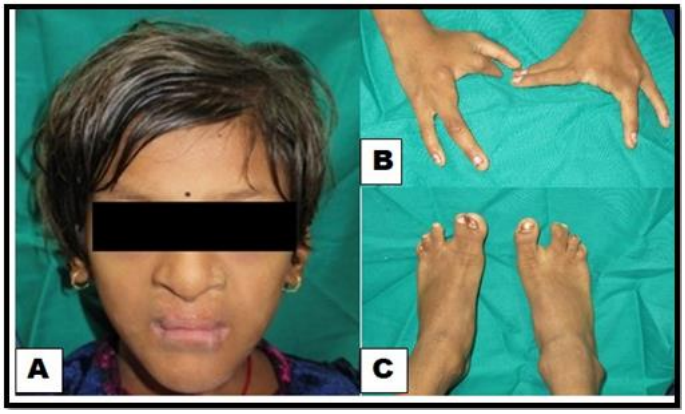

Figure 1: A- Patient with light pigmented hair, operated Bilateral cleft lip and palate, B- Upper limb, Split-hand

with missing central digits, C- Lower limb, syndactyly fusion of the digits.

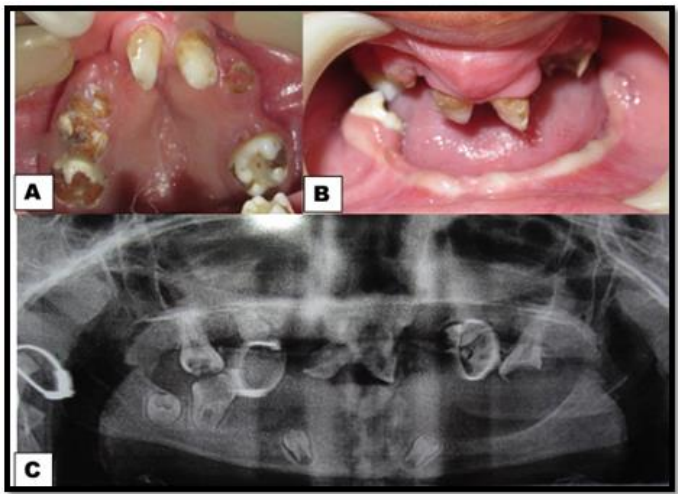

Figure 2: A- Maxilla representing carious 16, 54, 53, 63, 26, B - Mandible representing carious 46, C- OPG revealing the Unerupted impacted 14, 24, 47, 44, 34.

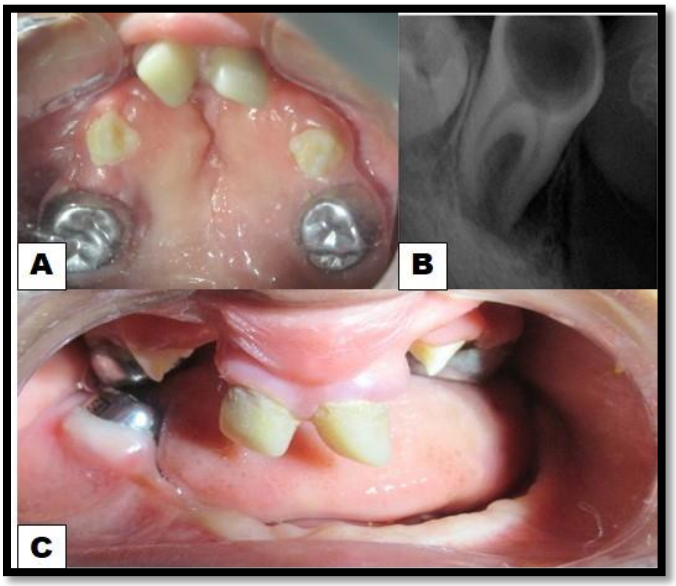

Figure 3: A- Stainless Steel crown cemented in 16, and 26, erupted 14, and 24, restored hypoplastic 11, and 21, B- IOPA revealing carious 46 with periapical radiolucency, C- Stainless steel crown cemented in 46.