PROSTHETIC MANAGEMENT OF PATIENT WITH PAPILLON-LEFEVRE SYNDROME: A CLINICAL REPORT

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ABSTRACT

Papillon Lefevre syndrome is a rare autosomal recessive genetic disorder, which is characterized by palmoplantar hyperkeratosis, with rapidly progressive periodontitis and premature loss of both deciduous and permanent teeth. The etiology of PLS is multifactorial with genetic, immunological, and microbial factors playing a role in etiopathogenesis. This is a case report of prosthodontic rehabilitation of a 14-year-old boy with Papillon-Leefevre syndrome.

Keywords: Papillon Lefevre syndrome, Palmoplantar hyperkeratosis, Periodontitis, Overlay prosthesis

INTRODUCTION

Papillon Lefevre syndrome (PLS) is a very rare autosomal recessive disorder characterized by palmoplantar hyperkeratosis and severe early onset periodontitis with premature loss of the primary and secondary dentitions [1]. This syndrome was first reported and described by two French physicians, Papillon and Lefevre in 1924[1].

The prevalence of PLS has been reported as 1 to 4 per million [2]. Parental consanguinity is demonstrated in between 20% and 40% of the cases [1].

From a dental standpoint, young patients with PLS have juvenile periodontitis, severe destruction of the alveolar bone as early as within two years of eruption in both primary and permanent dentitions. Associated features may include calcification of falx cerebri and choroid plexus, and retardation of somatic development[3-5]. Primary dentition is usually exfoliated prematurely by the age of 4 to 5 years [1,5]. Later, the third molars also undergo the same fate. Severe resorption of alveolar bone gives the teeth a ‘floating-in-air’ appearance on dental radiographs.

Conventional removable prosthodontics with or without osseointegrated dental implants can help to restore an efficient functional dentition. Psychological reassurance and regular follow-up will be the key to success in these complicated scenarios[7].

CASE REPORT

A 14-year old male patient reported to Dept. of Prosthodontics of Dr. R Ahmed Dental College & Hospital, Kolkata with chief complaint of unpleasant appearance, difficulty in chewing (Fig.1). Family history of patient revealed consanguineous marriage
of his parents. Patient gave a dental history of premature exfoliation of his teeth from childhood. Examination of patient showed Palmoplantar keratosis, with dry scaly keratotic plaques over the skin of his legs (Fig.2) along with multiple missing teeth and only 3rd molars remaining, associated with aggressive periodontitis (Fig.3). Radiographic examination revealed the classic presentation of “floating in air appearance” of remaining teeth (Fig.4).

Considering Muller De Van statement of “The preservation of that which remains is of utmost importance and not the meticulous replacement of that which has been lost”, to simplify the treatment plan and considering the age of patient and ongoing growth any extensive surgical options were avoided.

To restore function and esthetics, complete denture prosthesis overlaying the erupting 3rd molar was considered as the treatment option. Impression of maxillary arch was made with help of elastomeric impressions due to presence of tuberosity undercut and with zinc oxide eugenol impression paste for mandibular arch (Fig.5).

This was followed by taking Jaw relation of patient to determine vertical dimension. After teeth arrangement try-in was done (Fig.6). And phonetics, aesthetics were determined at this stage. Denture was delivered and patient was put on follow up at bimonthly interval. Patient was completely satisfied with aesthetics and function of denture (Fig.7). After 4 month of denture wearing tooth started to erupt in maxillary arch which was relieved adequately. As the patient is still in growing stages, Regular follow up of the patient need to be done to adjust and refine the denture in case need arises.
DISCUSSION

Dental surgeon is the first to diagnose Papillon-Lefevre because of severe periodontitis affecting the patient. This syndrome is inherited as an autosomal recessive trait with a prevalence of 1 to 4 cases per million\(^2\). PLS is characterized by marked destruction of the periodontium (periodontoclasia) of both dentitions with premature loss of teeth, marked palmar and plantar hyperkeratosis. The gene has been mapped to the long arm of chromosome 11. These patients are usually normal at birth with only reddening of palms and soles. Teeth erupt usually in normal sequence, position and time. At around one and an half to two years, marked gingivo-periodontal process develops with edema, bleeding, alveolar bone resorption and teeth mobility with consequent exfoliation\(^{1,5,6,7,8}\). The pathogenesis of this syndrome is still not understandable.

Etiology can be of Immunologic, genetic, and microbiologic factors. As a genetic factor cathepsin C gene is associated with Papillon-Lefevre syndrome \(^9,10,11\). The cathepsin C gene is seen in the epithelial regions commonly affected by Papillon-Lefevre syndrome, such as the palms, soles, knees, and keratinized oral gingiva\(^12\). In addition, it is expressed in various immune cells including polymorpho nuclear leukocytes and macrophages, as well as their precursors\(^13\). After exfoliation of all teeth, the soft tissues usually acquire the normal healthy appearance. The permanent dentitions usually start to appear at normal time, but just after 2 - 3 years, the gingivo-periodontal condition starts to deteriorate again. All permanent teeth usually exfoliate within a few years except for third molars which usually stay longer\(^{11,5,6}\). Peripheral blood neutrophil chemotaxis have been reported to be depressed. This decreased chemotaxis suggests that neutrophils may be important factor in periodontal destruction\(^{14-18}\).

No definitive treatment is available for prevention and management of periodontal destruction, although strict oral hygiene maintenance, scaling and root planning along with suitable antibiotic regimen may improve the situation. The treatment should be planned with a multidisciplinary team approach involving paediatricians, periodontists, dermatologists, prosthodontists and psychologists\(^19\). However, edentulous patients can adapt to removable prosthesis very quickly because of young age, better oral stereognostic and oral motor abilities\(^20\). The osseointegrated dental implants have revolutionized the possible treatment options, but long-term effects in these syndromic cases are still pending\(^19\). The Papillon lefevre syndrome can adversely affect growing children psychologically, socially and aesthetically. A multi-disciplinary approach may improve the prognosis and quality of life of these children. Thus, oral rehabilitation in such patients is a must.

Thus, prosthetic replacement in such patients is an age specific, speciality treatment involving initial replacement with complete or partial dentures and future consideration for an implant-supported prosthesis\(^21\). In the present case, prosthetic rehabilitation with an overlay prosthesis was considered as it would provide immediate satisfaction to the patient in terms of aesthetics and function.

CONCLUSION

Tooth supported over denture in addition to preserving the underlying tooth structure helps to restore form and function. Rehabilitating such a

Patient at a young age is important to help them adapt, maintain muscle tonicity and in overall development of stomatognathic system, so as to prepare them for more extensive treatment which may be desired in future. Follow-up at regular interval is important to see for any changes due to erupting teeth and modify the denture accordingly.

Conflict of Interest: Nil

REFERENCES


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