CASE REPORT

Prosthodontic Rehabilitation of Papillon-Lefèvre Syndrome

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ABSTRACT: Papillon-Lefèvre syndrome (PLS) is an autosomal recessive genetic disorder. It has a reported incidence of 1- 4 persons per million. This syndrome presents with dermatological and dental manifestations. It is usually diagnosed by the presence of palmar-plantar hyperkeratosis of the skin and premature exfoliation of the teeth. This case report describes preventive prosthetic treatment given to 17 year old female patient with PLS who had reported with old unaesthetic denture.

Key words: *Papillon-Lefèvre syndrome, Preventive Prosthodontics, Palmar-plantar hyperkeratosis*

Papillon-Lefèvre syndrome (PLS) is a familial disease, transmitted as an autosomal recessive disorder [1]. This syndrome was first reported in the literature by two French physicians Papillon and Lefèvre in 1924. It is a rare syndrome with an incidence rate of 1-4 person per million [2]. Recent research activities have found an association between the syndrome and cathepsin C gene mutation which leads to deficiency of cathepsin C enzymatic activity [3]. Parental consanguity is also attributed as one of the cause for this syndrome.

Patients affected by PLS present with palmar-plantar hyperkeratosis, periodontal pathology, early loss of dentition and increased susceptibility to infections [4].

Tooth loss in Papillon-Lefèvre syndrome is because of inflammation and destruction of the periodontal tissues. This syndrome affects both primary and permanent dentition; loss of teeth usually occurs in a sequence in which they have erupted. Pre-mature exfoliation of primary and permanent teeth, result in loss of most of the permanent teeth by 14-16 years of age. [5] The exact reason for severe periodontitis is however unclear. Saahan et al suggested that the destruction of periodontium may be due to imbalance in collagenolytic activity [5]. Page RC et al suggested that the reason for severe periodontitis would be due to defect in the development of cementum [6].

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The following case describes the prosthodontic management of Papillon-Lefèvre syndrome.

CASE REPORT

A 17 year old female patient reported to the Department of Prosthodontics, Indira Gandhi Institute of Dental Sciences, Sri Balaji Vidyapeeth, Puducherry, with the chief complaint of unaesthetic old denture. The patient gave a medical history which revealed that 4 years back she was diagnosed having Papillon–Lefèvre syndrome in the Department of Dermatology, Mahatma Gandhi Medical College and Research Institute, SBV. She was treated for the same with Isoretinoin, three times daily to improve the dermatological problem.

Patient gave a dental history of premature exfoliation of her teeth from her childhood. On enquiring the family history, patient said that her parents had a consanguineous marriage but her siblings are not affected by this disease.

Denture history revealed that the existing denture was delivered to the patient in a private clinic 5 years back. On inspecting the prosthesis, it was found that existing tooth was not involved in the treatment. Moreover calculus deposition was seen over the flanges of the upper and lower denture.

On general examination, the patient was moderately built with dry scaly thick keratotic plaques over the skin of her soles extending on the dorsal surfaces of hands and

legs (Fig-1).

On oral examination, the patient was partially edentulous, with only one tooth remaining (i.e) left mandibular third molar , 38. On examination of 38, (Fig-2) it was periodontally healthy with normal probing depth, which was confirmed by IOPA radiograph. (Fig-3). The radiograph revealed adequate crestal bone surrounding 38, with normal crown and root.

This clinical condition can be prosthetically managed by implant supported prosthesis and preventive prosthodontics. Muller De Van (1952) stated, "The preservation of that which remains is of utmost importance and not the meticulous replacement of that which has been lost" [7]. So we aimed at giving preventive prosthodontic treatment and planned a tooth supported overdenture to preserve the existing bone. As the patient could not afford, the option of implant supported prostheis was ruled out.

Preliminary impression was made with alginate (Tropicalgin, Zhermack,Italy) to make a diagnostic cast. Treatment planning was done to make a conventional upper complete denture and tooth supported lower overdenture.

An intentional root canal treatment (Fig-4) was performed for 38 in the Department of Conservative Dentistry and Endodontics, IGIDS. The tooth was reduced to 1 – 2 mm above the gingiva and the gutta percha was removed to about 6mm from the distal canal and the post space was prepared with peeso reamer. The crown preparation was also done to receive a metal coping with post.

Direct wax pattern was made (Type 1 wax, GC India) and was cast with nickel chromium alloy (BEGO, USA) (Fig-5). The cast metal coping with post was checked for the fit and then was cemented over 38 with type 1 luting cement (GC, India). Special tray was fabricated using the diagnostic cast for the final impression. Border molding was done with green stick compound (DPI, India). Upper final impression was made with Zinc oxide eugenol impression paste (DPI, India) and lower impression was made with light body material (Photosil, DPI, India) (Fig-6) after applying tray adhesive over the custom tray. Master cast (Goldstone, Asian chemicals, India) was made from the final impressions (Fig-7) and indexed. Self cure acrylic resin (DPI, India) was used for fabricating trial denture base. Bite rims were fabricated

Fig 1: Keratotic plaques on the dorsal surfaces of the hands and legs

Fig 2: Maxillary edentulous arch and mandibular arch with 38

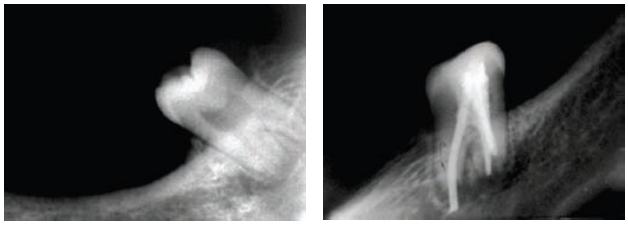

Fig 3 & 4: Pre-operative and post root canal treatment in 38

Fig 5: Direct wax pattern and cemented cast metal coping

Fig 6-8: Maxillary and mandibular final impressions, casts and dentures

and jaw relations were recorded. Teeth setting was done with cross- linked double layered acrylic teeth set (Acry-rock Ruthenium group, Italy). Try in was done and evaluated for esthetics, occlusion and phonetics. After try-in, patient's acceptance for the same was obtained. Then the denture was fabricated using heat cure denture base resin (DPI, India). After processing, laboratory remounting was done and occlusion was adjusted. Finally the denture was finished, polished and delivered to the patient.(Fig-8)

The patient was asked to wear the denture for first 24 hrs and report for follow up. Post insertion follow up was done after 24 hrs, 3 days, 1 week, 3 months and thereafter for every 6 months. Necessary adjustments were done during initial follow up to maintain good function, esthetics and occlusion. (Fig-9)

Patient was educated about the use and maintenance of the denture and the same was demonstrated to the patient.

DISCUSSION

Papillon-Lefèvre syndrome (PLS) is an autosomal recessive genetic disorder which gives a negative impact to the affected individuals both physically and mentally. Recent research in this field, has identified a genetic defect in PLS which is mapped in chromosome 11q14-q21, which involves mutation of cathepsin C.[8]

Taşlı PN et al advocated a treatment using dental pulp stem cell for cellular therapy of alveolar bone maintenance and other dental tissue abnormalities, observed in PLS [9].

Dental management of PLS patients is a multidisciplinary approach. The patients may present with partially or completely edentulous situation [10]. Existing dentition in partially edentulous patients may not be favorable because of the periodontal destruction. Rűdiger et al suggested proper and regular periodontal maintenance to lengthen the life span of existing teeth by following oral hygiene instructions, 0.06 % chlorhexidine digluconate jet irrigation and short- term use of metronidazole and amoxcillin with clavulinic acid [11].

As far as prosthetic options are considered, it ranges from removable partial dentures to implant supported fixed prosthesis. Shah et al treated a 3 year old child affected by PLS with telescopic dentures [12].

Fig 9: Pre-treatment and post-treatment photograph

The PLS case presented in this article had a good periodontium around 38. This gave a hope to proceed with an overdenture. Treatment in this case aims at providing proper function and esthetics as well as improve the self-confidence of the patient. Importantly, it will preserve the ridge for future prosthesis.

Vikhe DM et al treated an 18 year old boy affected by Papillon – Lefèvre syndrome by overdentures because the patient had two permanent molars in each arch with good periodontium which was modified by placing metal copings over it [13]. In case of fair prognosis of the existing teeth, transitional denture is fabricated with windows for permanent teeth.

Ahmadian L et al treated an edentulous 21 year old girl affected by PLS [14]. Treatment comprised of maxillary and mandibular fixed prosthesis supported by osseointegrated dental implant. The patient had no complication during 4 years of follow up period. Implant placement in PLS syndrome patients help to preserve the existing alveolar bone. In cases with severe resorption of the residual ridges, short implants have also been tried [15].

To summarize, Prosthetic management of Papillon-Lefèvre syndrome depends on the patient's existing dentition, health of the existing periodontium, amount of the remaining bone and cost factors. Partial edentulism in PLS patients with healthy periodontium of the existing teeth can be treated with cast partial dentures and implant supported fixed partial denture. If only few teeth are remaining with healthy periodontium overdentures are viable options. When there is unhealthy periodontium, dentures with windows are fabricated. Completely edentulous patients can be treated either by conventional dentures or implant supported dentures.

CONCLUSION

PLS is a condition in which patient suffers from dermatological and dental problems. Successful prosthodontic treatment of PLS patients must aim at restoration of proper function and esthetics. Prosthodontic rehabilitation of this patient improved her esthetics and restored her confidence level.

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