



## MANAGEMENT OF A YOUNG EDENTULOUS PATIENT HAVING PAPILLON LEFÈVRE SYNDROME – A CASE REPORT

1. Prof. Dilip Kumar Nath\* , MDS, HOD, Department of Prosthodontics, Chandra Dental College & Hospital, Safedabad, Barabanki, Uttar Pradesh, India. Pin : 225003 . E.Mail:[dilip\\_nath2006@yahoo.co.in](mailto:dilip_nath2006@yahoo.co.in)
2. Dr. Gautam Bagchi, MDS, Senior Lecturer, Department of Prosthodontics , Chandra Dental College & Hospital, Safedabad, Barabanki, Uttar Pradesh, India. Pin : 225003 . E.Mail: [gautambagchi79@yahoo.in](mailto:gautambagchi79@yahoo.in)
3. Madhuri Nath ( Final Year BDS Student ), Chandra Dental College & Hospital Safedabad, Barabanki, Uttar Pradesh, India. Pin : 225003 E.mail: [madhuri.saheli@gmail.com](mailto:madhuri.saheli@gmail.com)

### \*Corresponding Author :

Dr. Dilip Kumar Nath

9, Ashok Nagar , G.B. Marg ,Lucknow , UP . Pin : 226018, Phone : 0522 – 2636058, 📞 : 9838 646767

Email : [dilip\\_nath2006@yahoo.co.in](mailto:dilip_nath2006@yahoo.co.in)

### ABSTRACT

Papillon-Lefevre syndrome is a rare autosomal recessive disorder in which the frequently observed manifestations are palmoplantar keratinization and premature loss of both deciduous and permanent teeth. The palmoplantar keratoderma typically has its onset between the ages of 1 and 4 years and severe periodontitis starts at the age of 3 or 4 years. The deciduous teeth frequently fall out by about age five and most of the permanent teeth may also be lost by approximately age 17. In the present, a young, edentulous male of this syndrome having most the characteristic features was treated by modified Complete Denture prosthesis considering his young age and low socioeconomic status.

**Keywords:** Palmoplantar hyperkeratosis, Papillon-Lefevre syndrome, Complete denture prosthesis, Preservation of residual alveolar ridge.

### INTRODUCTION

Premature loss of teeth due to severe periodontitis and dermatological signs have been reported to be the characteristic features of Papillary Lefèvre syndrome, an autosomal recessive disorder. The immune system is also affected owing to decreased function of neutrophils, lymphocytes, or monocytes. An increased susceptibility to bacterial infection, leading to recurrent pyogenic infections of the skin and pyogenic liver abscess is another complication<sup>i</sup>.

The syndrome was first described by Papillon and Lefèvre in the year 1924<sup>ii</sup>. They reported a brother and a sister with palmoplantar hyperkeratosis associated with severe early-onset periodontitis and premature loss of primary and permanent teeth.

The parents are generally unaffected owing to its autosomal nature. Consanguinity is noted in approximately one third of cases. It is an extremely rare disease with 1 to 4 per million<sup>iii</sup>.

PLS usually manifests during the first 4 years of life with sharply demarcated hyperkeratosis,

more pronounced on the soles of feet and possibly extending to the dorsa of the hands and feet. Erythematous hyperkeratotic plaques may also be present at the elbows, knees, and trunk. The second major feature of PLS is severe periodontitis, which starts at the age 3 or 4 and affects both the deciduous and permanent teeth. The teeth erupt normally but fall soon and by the end of teen age patients are usually edentulous<sup>iii</sup>.

### Case Report

A 25-year-old, small built, unmarried, complete edentulous male patient from low socioeconomic background reported to the Prosthodontic department of Chandra Dental College for oral rehabilitation. The past dental history revealed that his deciduous teeth had erupted normally, but exfoliated gradually by the age of 6 years. Similarly, all his permanent teeth were also lost by the age of 20 despite having erupted



Fig 1: Symmetrical, well-demarcated, hyperkeratotic lesions of PLS

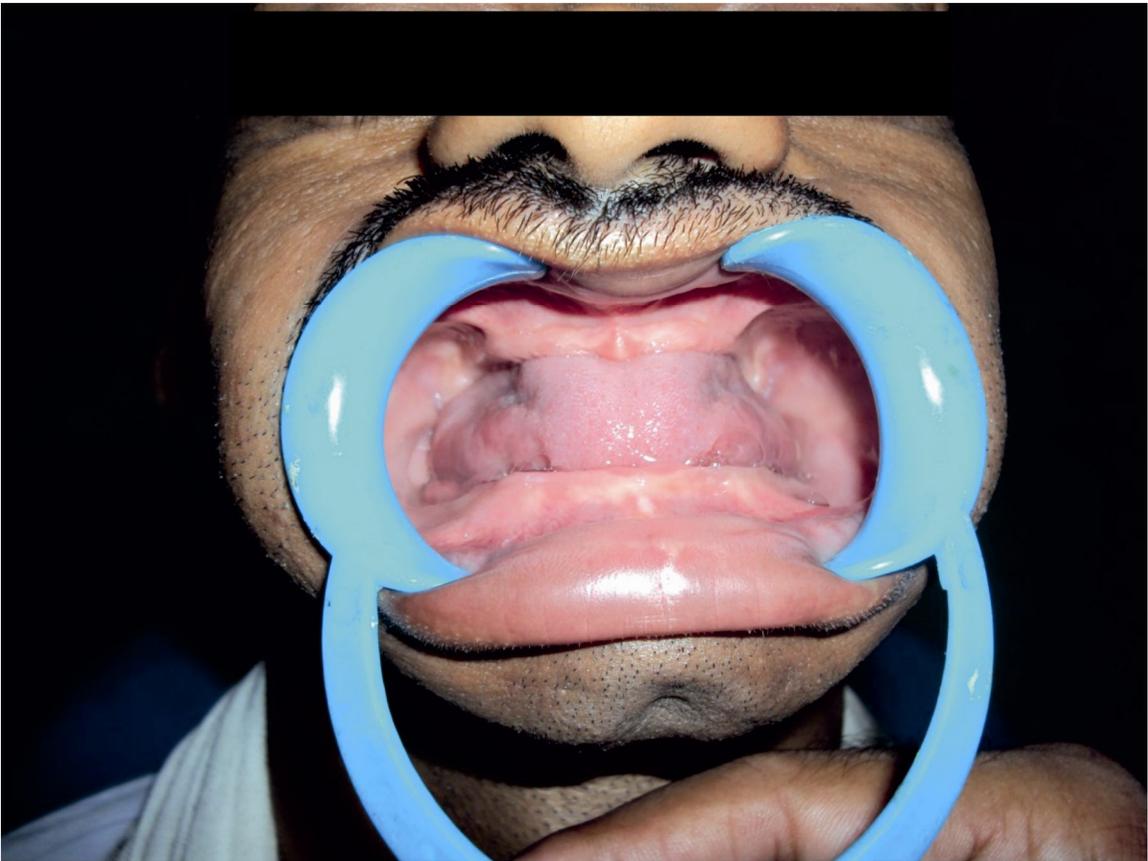
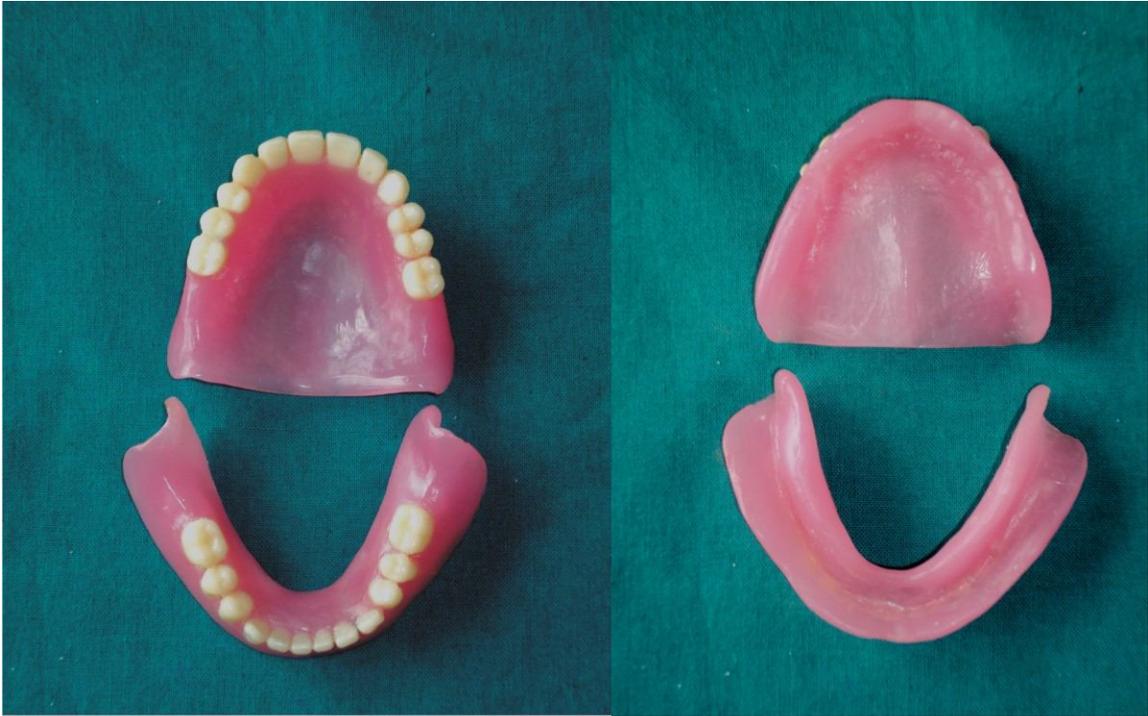


Fig 2: Small edentulous ridges



**Fig 3**

Fig 3: Modified Complete dentures with narrow posteriors and missing 2<sup>nd</sup> molars.



**Fig 4**

Fig 4: Patient before and after complete denture treatment.

normally. At the age of 2 years his parents had noticed the presence of rough skin on the plantar surface of his feet, with subsequent involvement of the palmar surface of the hands by the age of 3 years, with no relevant family history. The lesions became worse during

winters, with fissuring and bleeding from the fissures.

On examination, there were symmetrical, well-demarcated, keratotic and confluent plaque regions affecting the skin of the palms and soles which extended to the dorsal surface of the

finger joints. Well-circumscribed, psoriasiform, scaly plaques were also present on the elbows and knees bilaterally along with dystrophy and transverse grooving of the nails and excessive perspiration (hyperhidrosis).

The maxillary and mandibular edentulous ridges were smaller in size but normal in appearance. OPG of the patient showed absence of any tooth bud or root in either of the edentulous jaws. The skull X-ray showed no sign of intracranial calcification.

Laboratory data — Complete blood picture, liver function transaminase levels, total bilirubin, and alkaline phosphatase were also found to be normal.

Histopathology investigation could not be done as patient declined biopsy from the affected parts of palm & sole.

Diagnosis of Papillon-Lefèvre syndrome (PLS) was dependent mainly on physical signs.

The patient was advised for Complete Denture prosthesis considering young age, low socioeconomic status and financial constraints.

**As the patient was young and much of the residual alveolar ridge resorption already had taken place, certain modifications were deemed necessary during fabrication of Complete Dentures Prosthesis as follows :**

- Selective pressure impressions were made using ZnO impression paste<sup>iv</sup>.
- Vertical Dimension of occlusion was kept low i.e. ( IOG 5 mm )<sup>v,vi</sup>.
- Narrow posterior teeth were selected<sup>vii</sup>.
- Maxillary & mandibular - 2<sup>nd</sup> molars were omitted.
- Balanced occlusion was developed.
- Lesser use of prosthesis, avoiding hard foods and periodic checkup was advised.

#### **Follow Up**

The follow up was carried out at quarterly intervals upto 2 years. A set of preformed objective questions was asked and observations recorded in recall visits. The prosthesis was found to be functional and well-adapted. Prosthetic improvement in quality of life of the patient was found to be satisfactory.

#### **DISCUSSION**

The present case, dermatological and dental history strongly suggested the case to be Papillon Lefèvre syndrome (PLS) which included Palmoplantar hyperkeratosis associated with severe early-onset periodontitis and premature loss of primary and permanent teeth, hyperkeratosis of elbow & knee and nails were showing slight grooving and fissuring. However, eyelids, cheeks, labial commissures, legs, thighs, and axillae etc. were not affected. Although physical characteristics of the patient were typically suggestive of PLS, the laboratory findings of blood picture, liver functions - transaminase levels, total bilirubin, and alkaline phosphatase were not altered. Similar observations were made by Hegde and Reddy (2002)<sup>viii</sup> as also in agreement with Haneke<sup>ix</sup> who proposed the following three criteria to classify a case as PLS: (a) palmoplantar hyperkeratosis; (b) loss of primary and permanent teeth; and (c) autosomal recessive inheritance. The present case fulfilled all the three criteria with evidence of palmoplantar hyperkeratosis, loss of primary and permanent teeth as well as no sign of disease in parents indicating its recessive inheritance.

The prosthodontic management strategy for young edentulous patient must in general include the preservation of residual alveolar ridge preferably by implant prosthesis<sup>x,xi,xii</sup>. Literature is also available where removable partial denture has been used for prosthesis where some of the natural dentition is preserved<sup>xiii</sup>. However rehabilitation by using implant could not be employed due to financial constraints in present case and instead conventional complete denture was provided with modifications employing several measures e.g. use of selective pressure impressions<sup>xiv</sup>, low VRO (IOG 5 mm), narrow posterior teeth, missing 2<sup>nd</sup> molars, balanced occlusion, advised limited use of prosthesis and avoiding hard foods to reduce masticatory stresses in order to minimize residual ridge resorption, keeping in view a likely sequelae as long as the prosthesis is worn from the age of 25 years as the patient is in now.

#### **CONCLUSION**

A patient of PL syndrome from low socioeconomic background was managed by providing complete dentures with certain modifications like low VRO, narrow posterior teeth, missing 2<sup>nd</sup> molars, avoidance of hard foods and limited use of prosthesis keeping in

view the long-term prosthodontic needs of patient and compromised oral health status.

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