



Prosthodontic management of a young edentulous patient having Papillon–Lefèvre syndrome: A rare case report

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Abstract:

Papillon–Lefevre syndrome is a rare autosomal recessive disorder, in which the clinical manifestations are periodontitis which results in premature loss of both deciduous and permanent teeth and palmoplantar keratinization. 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 case, 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.

Key words: Keratoderma, Papillon–Lefevre syndrome, periodontitis

Papillon–Lefevre syndrome (PLS) is an extremely rare disorder with a prevalence of 1–4/million.^[1] It was first described by 2 French physician Papilla and Lefèvre in 1924.^[2] Consanguinity is evident in approximately 1/3 of cases.^[3] There is no sex and racial predominance. PLS usually manifests during the first 4 years of life with hyperkeratosis, more pronounced on the soles of feet. The second major feature of PLS is severe periodontitis, which affects both the deciduous and permanent teeth. The teeth erupt normally but fall soon and by the end of teenage patients is usually edentulous.^[4]

CASE REPORT

A 25-year-old, unmarried, complete edentulous male patient from low socioeconomic background reported to the department of prosthodontics. The past dental history revealed that his deciduous teeth and permanent teeth had erupted normally and there was the premature shedding of deciduous teeth by 5 years. Similarly, all his permanent teeth were also lost by the age of 20 years. There was a history of consanguineous parents. It was noticed that patient was socially demoralized due to loss of teeth and poor esthetics because of sunken cheek [Figures 1 and 2].

At the age of 12 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 13 years. On examination, there were no other associated problems except for the burning sensation of the palms and soles. Skin and hair were normal except mild hypohydrosis such as dry areas on palms, soles, and even extended on the fingers.

Extraoral examination revealed that the patient had poor esthetics, unsupported oral musculature leading to sunken cheeks [Figure 3].

Intraoral examination revealed that the maxillary and mandibular edentulous ridges were smaller in size but normal in appearance.

Laboratory report – Complete blood picture, liver function transaminase levels, total bilirubin, and alkaline phosphatase were normal.

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Figure 1: Small edentulous ridges



Figure 3: Extraoral examination

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

Diagnosis of PLS was based mainly on clinical signs.

The patient was advised for complete denture prosthesis with cheek plumper considering young age, low socioeconomic status, and financial constraints. All the steps for conventional complete denture were completed till try-in stage. At the try-in stage, the template for cheek plumper was fabricated with the help of impression compound. Impression compound was molded and placed over the buccal flange of the denture base [Figure 4]. Border movements were done so that the compound was well adapted. Movements were repeated till the cheeks gained required fullness. Now, the cheek plumper made of impression compound were separated from waxed up maxillary denture bases. Denture flasking and dewaxing procedures were finished separately for the final denture and cheek plumpers. The resultant mold space was then packed with heat-polymerizing acrylic material, and curing procedures were completed. After curing, the cured final prosthesis and maxillary cheek plumpers were retrieved. In case of the mandibular denture, denture flasking and dewaxing procedure were done. After curing, mandibular denture with attached cheek plumper was retrieved [Figures 5-7]. In mandibular denture, cheek plumpers were not separated as it provides more weight which helps in the stability of denture in case of resorbed ridge.



Figure 2: Hypohidrotic dermal lesion



Figure 4: Molded impression compound placed over buccal flange of the denture base

Clinical magnets were not affordable for the patient, so push button attachments were used to attach cheek plumper with denture base. Two 2-mm deep and 5-mm diameter holes were made on the posterior flange of the maxillary denture base and the corresponding area of cheek plumper also. The female part of the push button was attached to the denture base, and the male part of push button was attached to the cheek plumper with the help of autopolymerizing resin [Figures 8 and 9]. Patient was completely satisfied with aesthetics and function of denture [Figure 10]. Postoperative instruction was given to patient regarding the attachment of plumper as well as maintaining the cleanliness and hygiene of complete denture-plumper complex.

DISCUSSION

PLS is an extremely rare genetic disorder. PLS is characterized by the development of dry, scaly patches on the skin of the palms and the soles (palmar-plantar hyperkeratosis) in association with severe inflammation and degeneration of the structures surrounding and supporting the teeth (periodontium). The deciduous teeth frequently become loose and fall out by about age five. Without treatment, most

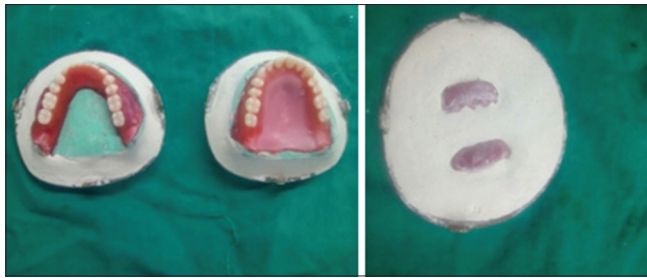


Figure 5: Flasking of denture and cheek plumper

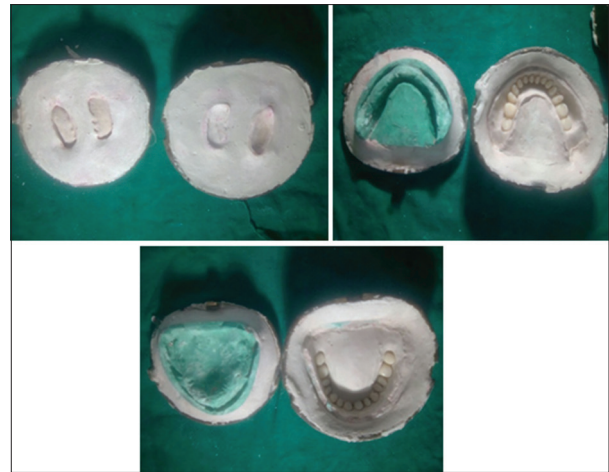


Figure 6: Dewaxing of denture



Figure 7: Finished and polished denture with cheek plumper



Figure 8: Female part of pushbutton attached to denture base and male part attached to acrylic cheek plumper



Figure 9: Cheek plumper attached to denture base

of the permanent teeth may also be lost by approximately age 17. Additional symptoms and findings associated with PLS may include frequent pus-producing (pyogenic) skin infections,^[5] abnormalities of the nails (nail dystrophy), and excessive perspiration (hyperhidrosis). PLS is transmitted as an autosomal recessive trait. It results from changes (alterations) of the CTSC gene that regulates the production of an enzyme known as cathepsin C.^[6] In today's world, denture esthetics is confined not only to selection of the teeth based on factors such as form, shape, color, arrangement, instead, it is more about harmonization between the artificial and natural tissues. Loss of teeth in the posterior region results in loss of cheek support due to which cheek tend to move medially to meet laterally expanding tongue. Rectifications of drooping of cheeks can be done



Figure 10: Postoperative photograph

by different methods such as reconstructive plastic surgery, injecting the botulinum toxin in the facial muscles, and different type of prosthesis.^[7] Conventional cheek plumpers which are a single-unit appliance with extensions on either side of the posterior flange of denture base leads to muscle fatigue and decreased retention.^[8] Therefore, detachable cheek plumpers provide an advantage to detach the cheek plumpers if they lead to muscle fatigue on long-term use. There are different attachment to fabricate cheek plumper prostheses such as magnet, push button, buccal tube, and

Co-Cr ball attachment. Magnetic attachment exhibits poor corrosion resistance and loss of magnetic property over time. Push button attachment has many advantages such as low maintenance cost, ease of replacement, cleaning, and better patient compliance. Clinical magnets being expensive, push button attachments are the most affordable means to attach cheek plumper to the denture.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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