

Rehabilitation of Papillon- Lefevre Syndrome Patient: A Case Report

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Abstract

Papillon-lefevre syndrome (PLS) is a rare Genodermatosis of autosomal recessive type characterised by diffuse palmoplantar keratoderma and precocious aggressive periodontitis, leading to premature loss of deciduous and permanent dentition at a very young age.

Papillon-Lefevre Syndrome is a devastating disease process characterized by rapid destruction of the dento-alveolar complex. In this paper, we describe a case report of a patient who had presented with severe periodontitis due to which extraction of teeth was advised in him, except the extraction of partially erupted third molars to preserve bone. Complete dentures were fabricated to accommodate the cusps of partially erupted third molars to aid in denture retention.

The success of complete dentures depends on a correct diagnosis, detailed treatment planning, and execution of fabrication procedures. A precise diagnosis and work plan can be made only after gaining insight into the patient's general health and detailed extra-oral and intraoral

examination. The patient's cooperation towards the treatment also plays a major role in success.

The aim of this clinical case report is to describe the steps involved in prosthetic rehabilitation of patient with papillon-lefevre syndrome.

Keywords: aggressive periodontitis, cathepsin-C gene, keratotic lesion, fluoride varnish.

Introduction

Papillon- Lefevre Syndrome is an inherited disorder characterised by cutaneous and oral manifestations. This disease was first explained in 1924 as a rare autosomal recessive genetic disorder by two French physicians Papillon and Lefevre.

Occurrence of this disorder is about one to four cases per million ^[1]. The exact immunologic abnormality is unknown and the disease is mainly associated with decreased neutrophil activity ^[2] as well as mutation in the Cathepsin C gene. The characteristic cutaneous lesions comprise of keratotic lesions of palmar and planter surfaces, these changes usually appear before the age of 4

years^[3, 4]. The oral lesions are characterised by aggressive periodontitis, resulting in a severe destruction of alveolar bone involving both deciduous and permanent dentitions^[5]. Conventional periodontal treatment usually fails to arrest the rapid progression of periodontitis and severe loss of alveolar bone is usually the result^[6]. Due to rapid bone loss, mobility and pathological migration occurs, which results in the loss of entire dentition. Prophylactic extractions of all permanent teeth has been considered as the treatment of choice to preserve the remaining supporting bone^[7].

Since both deciduous and permanent dentitions are affected, the patients become edentulous over a period of time, resulting in use of prosthesis at their teenage. Prosthetic rehabilitation of such patient can be done with either removable complete denture or implant supported overdenture.

Case Report

An 18 year old male patient presented to the department of prosthodontics, crown and bridge, aesthetic and implantology, Army College of Dental Sciences, Secunderabad with the chief complaint of missing teeth, loose teeth and difficulty in eating. Past dental history revealed a normal eruption of deciduous dentition. However, all the deciduous teeth had been exfoliated by the age of 4-5 years.

He gave a medical history of being diagnosed as suffering with papillon-levefre syndrome and was undergoing dermatologic treatment for the hyperkeratosis of his palms and soles.

On extra oral examination [fig 3, 4], well-demarcated, rough, hyperkeratotic, scaly lesions were found on palms and soles with fragile nails. Deepened mento-labial sulcus was present due to reduced vertical dimension of occlusion.

On intraoral examination, the teeth present were 13, 23, 27, 37, 35 with erupting 38, 48, 28 (only small portion of cusp visible clinically), the oral hygiene was poor and showed considerable periodontal involvement.

Radiographic examination revealed generalised advanced horizontal bone loss involving both maxilla and mandible. Extraction of all the teeth was planned except the erupting third molars.

Following a detailed complete examination and careful evaluation of the objective parameters of the patient i.e. age, aesthetic, functional demands and affordability, conventional complete dentures were planned.

Procedure

Extraction of teeth was performed and the healing was uneventful [fig 1, 2]. The Primary impression was made with impression compound and the primary cast was poured using dental plaster. A special tray with spacer was fabricated with an extension to include the erupting cusps of third molars. Border moulding followed by secondary impression using zinc oxide eugenol paste was made [fig 5, 6] and the master cast was poured using dental stone [fig 7]. Denture base plate and occlusal rims were constructed.

Facebow record taken and mounted on semi adjustable Hanau articulator after assessing the vertical jaw relation [fig 8, 9, 10, 11].

Complete maxillary and mandibular dentures were fabricated with extensions to include the erupting third molar cusps under the denture [fig 12, 13]. The denture provided adequate lip and cheek support and had excellent retention and the cusps of third molars included under denture provided adequate retention. Before delivery of denture, fluoride varnish was applied over the erupting cusps of third molars as a protective coating to prevent caries and also protection from demineralization and erosion. Denture related instructions were given and

additional oral hygiene instructions were also provided to minimise the chances of caries or inflammation of gingiva around erupting third molar.

The patient was satisfied with the complete dentures as both the functional and aesthetic demands were fulfilled [fig 14]. The patient was on review until the complete eruption of third molars was observed, thus aiding in modification of the fabricated denture.

Discussion

The patient mentioned in our case report presented with pre-pubertal periodontal destruction and concomitant palmar-plantar hyperkeratosis and hence was diagnosed as suffering with Papillon-Lefevre Syndrome. Numerous etiologic factors have been identified and have a pivotal role to play in the development and progression of PLS. A major role is played by the mutation of the cathepsin-C gene that is expressed within the ordinarily affected epithelial regions like the palms, soles and knees and the keratinized oral gingiva. It's additionally expressed at high levels in numerous immune cells, together with polymorph nuclear leukocytes, macrophages and their precursors. The precise reason behind the periodontal disease in PLS has not been found, however it's been attributed to lower neutrophil phagocytosis, bacterial infection and impaired reactivity to T- and B-cell mitogens. Actinomyces actinomycetemcomitans, inflicting periodontal damage and alterations within the polymorph nuclear leukocyte functions, are postulated as being a probable pathogenic mechanism. Gingival infection, abscess formation, loss of alveolar bone and destruction of periodontal ligament are the main cause for the unnatural early exfoliation of teeth.

It starts affecting the individual during childhood and poses both physical and psychological challenges. It is well established that tooth extraction is followed by a

reduction of the buccolingual as well as apicocoronal dimension of the alveolar ridge at the edentulous site^[8,9].

Early extractions play a role in preservation of alveolar bone if periodontitis cannot be controlled^[10]. At the same time, a cost/benefit evaluation should be carried out to rationalize the prophylactic removal of third molars, which should only be indicated with the aim of preventing cases that involve pathological processes, like root resorption or caries in second molars, cysts and pericoronitis.

Systematic reviews report that there is no evidence to support or refuse prophylactic removal of asymptomatic impacted third molars, even in adults^[11, 12]. No long term studies exist which validate the welfare to the patient either of early removal or of deliberate retention of these teeth.^[13]

Therefore in this case third molars were not advised for extraction for the following reasons:

- Incomplete eruption
- Partial clinical visibility of cusps of 28, 38 and 48
- Absence of inflammation
- Absence of periodontal pockets around erupted third molar cusps
- Avoiding surgical removal of third molars
- Extension of complete dentures to help aid in retention.

A decision to retain the third molars requires periodic monitoring involving clinical examinations and radiographs.

Proper oral hygiene instructions have to be meticulously followed along with application of fluoride varnish over the erupted cuspal regions and relining of the denture if required with time.

References

1. Gorlin RJ, Sedano HD, Anderson VE. The syndrome of palmar-plantar hyperkeratosis and premature periodontal destruction of the teeth. *J Pediatr.* 1964; 65:895–898.
2. Van Dyke TE, Taubman MA, Ebersole JL, et al. The Papillon-Lefevre Syndrome: Neutrophil dysfunction with severe periodontal disease. *Clin Immunol Immunopathol.* 1984;31:419–429.
3. Bach, J.N., Levan, N.E., 1968. Papillon–Lefevre syndrome. *Arch. Dermatol.* 97, 154–158.
4. Yagmur, A., Yilmaz, G., Ertan, U., IKizoglu, E., Ozkasap, S., Karacan, C., 2004. Papillon Lefevre syndrome: a case report. *Int. Pediatr.* 19, 224–225.
5. Rajendran, R. *Shafer’s textbook of oral pathology.* Elsevier India; 2009.
6. Rateitschak-Pluss EM, Schroeder HE. History of periodontitis in a child with Papillon-Lefevre syndrome. A case report. *J Periodontol.* 1984; 55:35–46.
7. Machtei EE, Zubrey Y, Ben Yehuda A, et al. Proximal bone loss adjacent to periodontally “hopeless” teeth with and without extraction. *J Periodontol.* 1989; 60:512–515.
8. Lekovic V, Camargo PM, Klokkevold PR, et al. Preservation of alveolar bone in extraction sockets using bioabsorbable membranes. *J Periodontol* 1998; 69: 1044–1049.
9. Schropp L, Wenzel A, Kostopoulos L, et al. Bone healing and soft tissue contour changes following single-tooth extraction: a clinical and radiographic 12-month prospective study. *Int J Periodontics Restorative Dent* 2003; 23: 313–323.
10. Eick S, Puklo M, Adamowicz K. Lack of cathelicidin processing in Papillon-Lefevre syndrome patients reveals essential role of LL-37 in periodontal homeostasis. *Orphanet J Rare Dis.* 2014; 9:148.
11. Costa MG, Pazzini CA, Pantuzo MC, Jorge ML, Marques LS. Is there justification for prophylactic extraction of third molars? A systematic review. *Braz Oral Res* 2013; 27(2):183-8.
12. Mettes TG, Ghaemini H, Nienhuijs ME, Perry J, Van der Sanden WJ, Plasschaert AJ. Surgical removal versus retention for the management of asymptomatic impacted wisdom teeth. *Cochrane Database Syst Rev.* 2012 Jun 13; 6:CD003879.
13. Raymond P. White, William R. Proffit. Evaluation and management of asymptomatic third molars: Lack of symptoms does not equate to lack of pathology. *Am J Orthod Dentofacial Orthop* 2011; 140:10-7.

Legends Figure



Fig 1: Pre-op occlusal view showing erupting 28, 38 and 48 cusps.



Fig 2: Pre-op (frontal view)



Fig 3: External features



Fig 4: External features

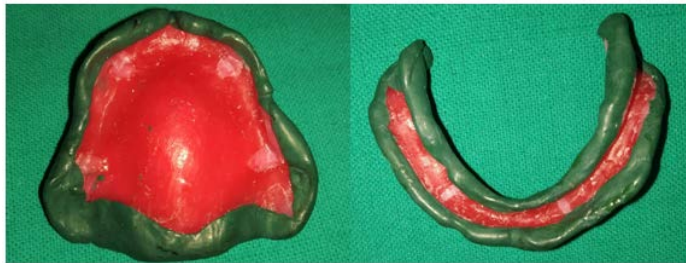


Fig 5: Border moulding of maxillary and mandibular arch



Fig 6: Secondary impression

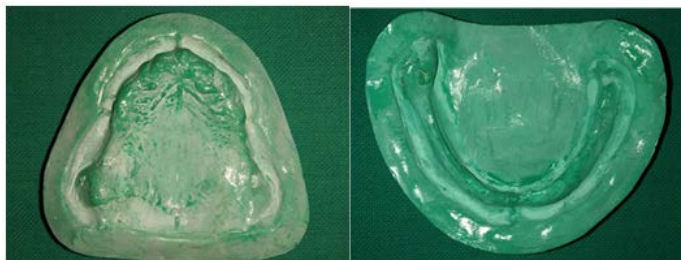


Fig 7: Maxillary Master cast Mandibular master cast

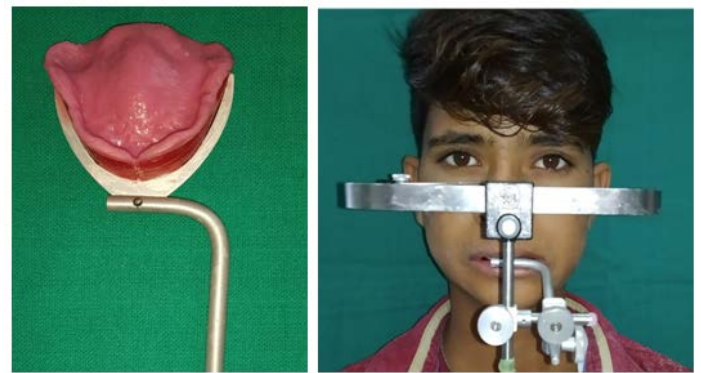


Fig 8: Face bow

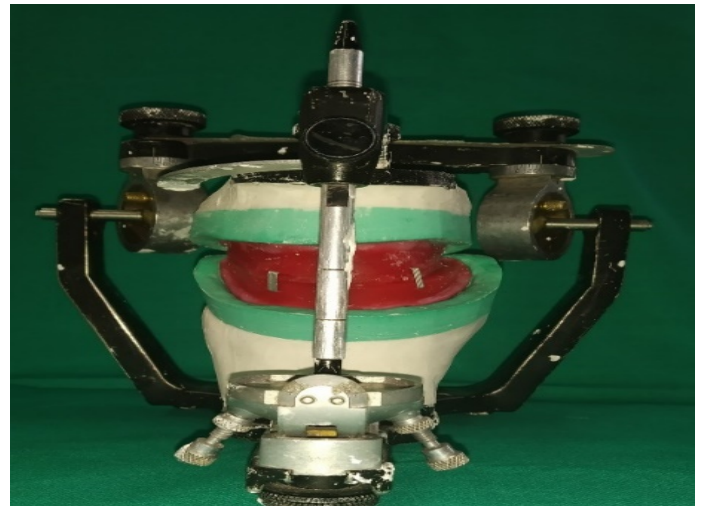


Fig 9: Mounting of master cast



Fig 10: Anterior teeth setting



Fig 11: Try-in



Fig 12: Final denture



Fig 13: Pre-op and post-op showing the improvement in the mento-labial sulcus



Fig 14: Final insertion.