DOI: 10.5958/2319-5886.2015.00135.6

International Journal of Medical Research & Health Sciences

www.ijmrhs.com Volume 4 Issue 3 Coden: IJMRHS Copyright @2015 ISSN: 2319-5886

Received: 15th Feb 2015 Revised: 10th Mar 2015 Accepted: 28th Apr 2015

Case report

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

Sushen Sharma[1], *Akhil Mittal[1], TK giri[2], Bhaskar Sengupta[3], Vishal Jain[4]

[1]PG student, Department of Prosthodontics, [2]Professor, [3]Professor & HOD, Department of Prosthodontics, Principal,[4]PG student, Department of Paedodontics & Preventive Dentistry, DR R Ahmed Dental College & Hospital, Kolkata, India.

*Corresponding author email: akhilmittal1988@gmail.com

ABSTRACT

Papillon Lefevre syndrome is a rare autosomal recessive genetic disorder, which is characterized by palmar-plantar 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-Lefevre syndrome.

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

INTRODUCTION

Papillone 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]. After exfoliation, the inflammation subsides and the gingiva appears healthy. With the eruption of permanent dentition, the whole process of gingivitis and periodontitis is repeated and there is subsequent premature exfoliation of the permanent teeth by the age of 14-15 years [1,5,6]. 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 14year 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[8] "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, compete 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 reline the denture in case need arises.

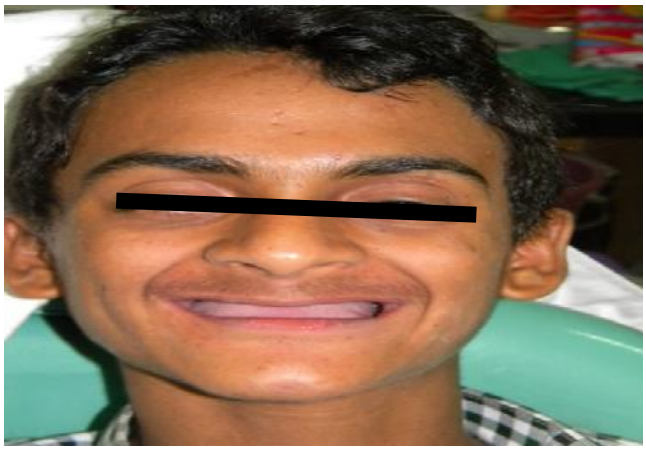

Fig 1: Pre-operative appearance

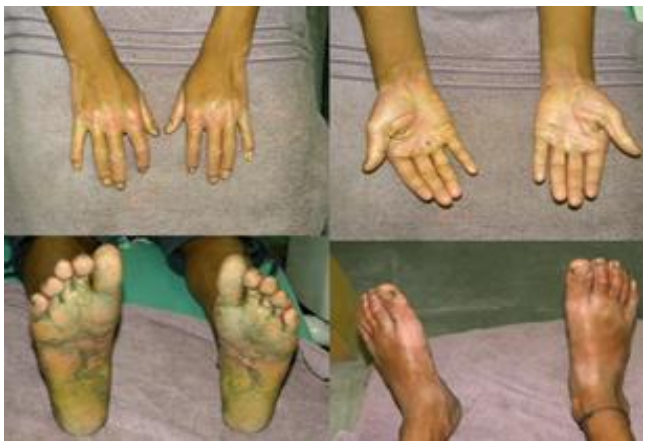

Fig 2: Hand and feet showing Palmoplantar keratosis

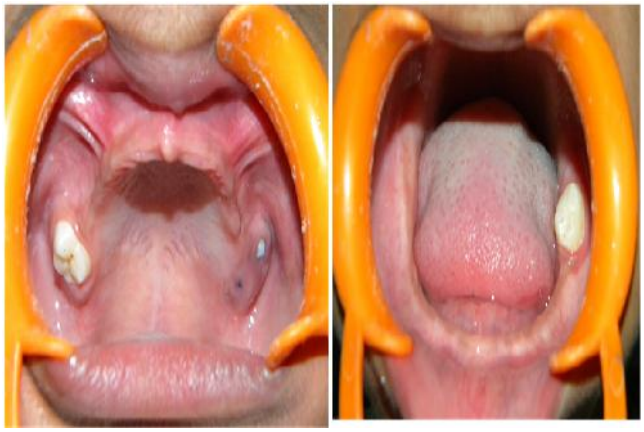

Fig 3: Intra-oral view of maxilla and mandible

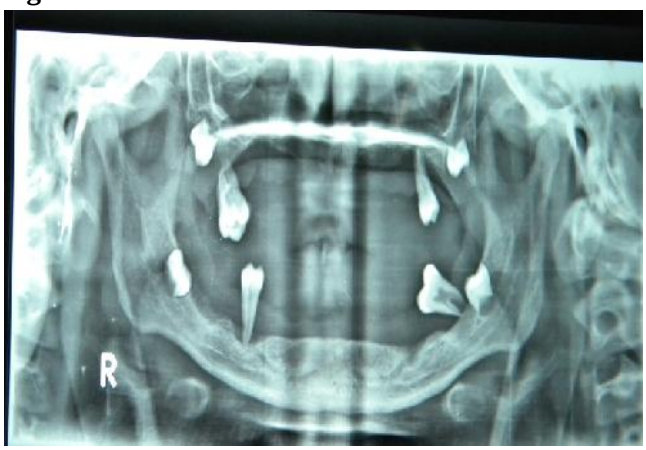

Fig 4: OPG showing floating in air appearance

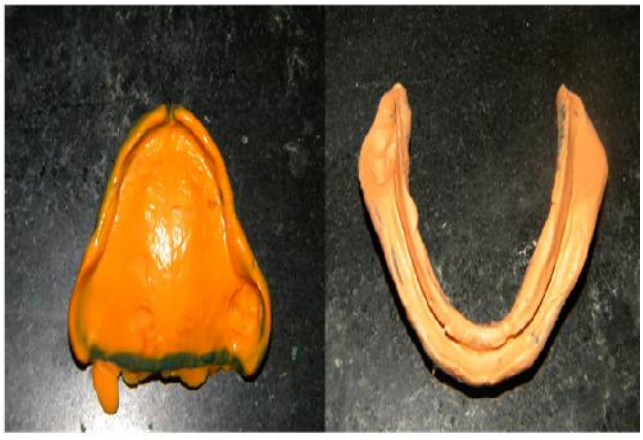

Fig 5: Final impression of maxilla and mandible

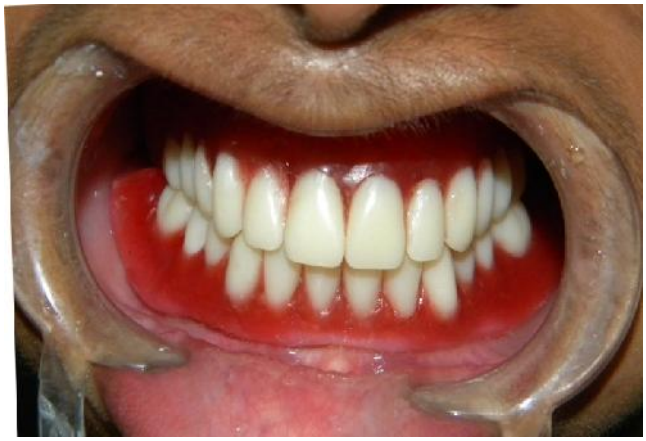

Fig 6: Try-in denture

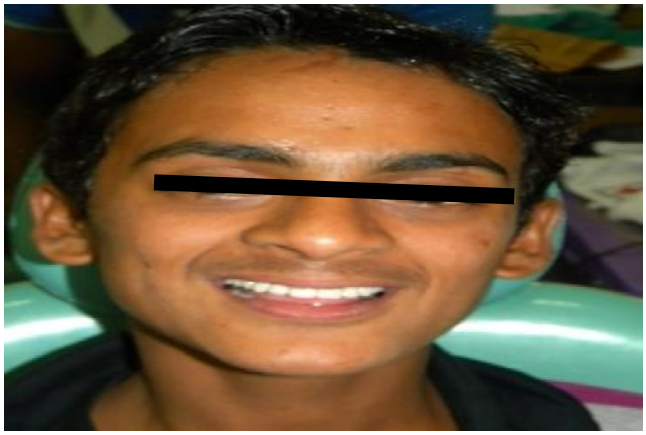

Fig 7: Final denture

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[1,5,6]. 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

1. Papillon MM, Lefèvre P. Deux cas de Keratodermie Palmaire Et Plantaire Symétrique Familiale (maladie de Meleda) Chez Le Frere Et La Soeur. Coexistence dans les deus cas alterations dentaires grabes. Bulletin de la Soceite Francaise de Dermatologie et de Syphiligraphie 1924; 31, 82–87.
2. French D, Scott H, Overall CM. Papillone Lefevre syndrome associated early onset periodontitis: a review and case study. J Can Dent Assoc. 1995; 61:432-38.
3. Gorlin RJ, Cohen MM, Levin LS. Syndromes of the Head and Neck, 3rd edn.Oxford:Oxford University Pres; 1990: 853–55.
4. Hall RK (ed.). Paediatric Orofacial Medicine and Pathology, 1st edn. London: Chapman & HallMedical; 1994;Vol/issue: page no
5. Kressin S, Herforth A, Preis S, Wahn V, Lenard HG. Papillon – Lefèvre syndrome–successful treatment with a combination of retinoid and concurrent systematic periodontal therapy: case reports. Quintessence International 1995; 26:795–03.
6. Wara-Aswapati N, Lertsirivorakul J, Nagasawa T, Kawashima Y, Ishikawa I. Papillon–Lefevre syndrome: serum immunoglobulins G (IgG) subclass antibody response to periodontopathic bacteria. A case report. Journal of Periodontology 2001; 72: 1747–54.
7. Subramanium P, Mathew S, Gupta KK. Papillon Lefevre syndrome: a case report. J Indian Soc Paedo Prev Dent 2008; 26:171-4.
8. DeVan M. The nature of the partial denture foundation: suggestions for its preservation. J Prosthet Dent. 1952;2:210-18.
9. Hart T C, Hart PS, Bowden DW, et al. Mutation of the cathepsin C gene are responsible for Papillon–Lefèvre syndrome. Journal of Medical Genetics 1999; 36: 881– 87.
10. Toomes C, James J, Wood AJ et al. Loss-of-function mutations in the cathepsin C gene result in periodontal disease and palmoplantar keratosis. Nature Genetics 1999; 23: 421–24.
11. Hart PS, Zhang Y, Firatli E, et al. Identification of cathepsin C mutations in ethnically diverse Papillon– Lefèvre syndrome patients. Journal of Medical Genetics 2000; 37: 927–32.
12. Cagli NA, Hakki SS, Dursun R, et al. Clinical, genetic, and biochemical findings in two siblings with PapilloneLefe`vre syndrome. J Periodontol. 2005; 76: 2322.
13. Sollecito TP, Sullivan KE, Pinto A, et al. Systemic conditions associated with periodontitis in childhood and adolescence (A review of diagnostic possibilities). Med Oral Patol Oral Cir Bucal. 2005; 10:142.
14. D'Angelo A, Margiotta V, Ammatuma P, Sammartano F. Treatment of prepubertal periodontitis. A case report and discussion. Journal of Clinical Periodontology 1992; 19: 214–19.
15. Brown RS, Hays G, Flaitz CM, O'Neill PA, Abramovitch K, White RR. A possible late onset variation of Papillon–Lefèvre syndrome: report of 3 cases. Journal of Periodontology 1993; 64: 379–86.
16. Tinanoff N, Tempro P, Maderazo EG. Dental treatment of Papillon–Lefèvre syndrome: 15 year followup. Journal of Clinical Periodontology 1995; 22: 609–12.
17. FQratlQ E, Gurel N, Efeoglu A, Badur S. Clinical and immunological findings in 2 siblings with Papillon–Lefèvre syndrome. Journal of Periodontology 1996; 67: 1210– 15.
18. Ghaffer KA, Zahran FM, Fahmy HM, Brown RS. Papillon–Lefèvre syndrome. Neutrophil function in 15 cases from 4 families in Egypt. Oral Surgery, Oral Medicine, Oral Pathology, Oral Radiology and Endodontics 1999; 88: 320– 25.
19. Etoz OA, Ulu M, Kesim B. Treatment of patient with Papillon Lefevre syndrome with short dental implants: a case report. Implant Dent 2010; 19:394-9.
20. Ahmed B, Mirza KM, Mehmood A, Hussain M, Yazdanie N. Oral stereognostic ability in

hypodontia patients. Pak J Med Research 2010; 49:14-7
21. Varri S, Chukka RS, Tadepalli A, Kiran R Prosthetic Rehabilitation For A Case Of Papillon-lefevre Syndrome: Indian Journal of Dental Sciences. March 2014, 1(6):88-89