PROSTHETIC REHABILITATION OF A PATIENT WITH PAPILLON LEFEVRE SYNDROME

[1]AQSA NADEEM, [2]HINA NAEEM, [3]HINA ZAFAR RAJA

ABSTRACT

A 16 year old male patient presented in Institute of Dentistry, CMH Lahore Medical College on 17th August, 2015. Presenting complaints were loose teeth, bleeding gums and inability to eat. On general examination patient appeared mentally stable. He had poor general health, weak built and belonged to a compromised socioeconomic background. He had yellowish keratinized plaques over the skin of his palms and soles. (Fig 1). Extra oral examination revealed normal hair and nails and a tapering face form. Intraoral presentation included heavy plaque deposits, halitosis, bleeding gums and pus exudates from pockets with grade 3 mobility. He was diagnosed with Papillon Lefevre Disease. After dental extractions, complete maxillary and mandibular dentures were planned for the patient. On denture insertion patient was satisfied. A follow up was done at 3 monthly interval. At the end of first year, it was observed that there was minimal ridge resorption and denture retention was moderate with adequate chewing ability. Patient did not have any complaint with denture. However, after two years the complete dentures were remade to improve the retention and stability of dentures. At completion of puberty, dental implants are planned to improve the quality of life for that individual. It was concluded that complete denture is a suitable prosthesis for Papillon Lefevre syndrome for restoration of mastication, phonetics and esthetics. It may need constant revisions depending on the stage of the disease and age of the patient.

Key Words: *Complete Denture, Papillon-Lefevre Disease, Periodontitis*

INTRODUCTION

Papillon Lefevre Syndrome is an autosomal recessive disorder.[1] Clinical presentation include palmoplantar keratoderma with a rapidly progressive periodontal disease.[2] This may lead to premature loss of both the deciduous and permanent dentitions.[2] Characteristic radiographic features include generalized horizontal and vertical bone loss with a classical "floating in air" appearance. On general examination patient may have intra cerebral calcifications, arachnodactyly, increased susceptibility to infections and mental retardation.[3] The prevalence of this disease is 1 to 4 per million people. It does not have any sexual and racial predominance.[3] In a patient with Papillon Lefevre syndrome there is mutation in Cathepsin C gene. It is mainly expressed in epithelial regions including palms, soles and keratinized oral gingival.[4] In addition, there is impairment of neutrophil serine proteinases and natural killer cell cytotoxic function.This leads to degranulation of localized polymorphonuclear cells, inactivation of phagocytic cells and T lymphocytes.[3] There is reduced response to A.actinomycetemcomitans and Staphylococcus species. Thus decreased immune response will increase the liability to infection.[4]

[1] Aqsa Nadeem, PGR Prosthodontics, Institute of Dentistry, CMH Medical College, Lahore Cantt, Cell: 0092-302-8425844 Email: axanadeem@live.com

[2] Hina Naeem, PGR Prosthodontics, Institute of Dentistry, CMH Lahore Medical College, Cell: 0092-336-8837760 Email: rockstone_402@rocketmail.com

[3] Hina Zafar Raja, Professor Prosthodontics, Institute of Dentistry CMH Medical College, Lahore Cantt, Cell: 0092-334-4222341 Email: hinazafarraja@gmail.com

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CASE REPORT

A 16 year old male patient presented in Institute of Dentistry, C.M.H Lahore Medical College. Chief complain were inability to eat, loose teeth and bleeding gums. Past dental history revealed normal eruption of deciduous dentition. However, all the deciduous teeth had been exfoliated around the age of 4-5 years. Past medical history revealed continuous flaking and scaling of skin. Symptoms became worse during winters due to dryness and superimposed infections. There was no relevant family history.

On general physical examination patient appeared mentally stable. He had poor general health, weak built and belonged to a compromised socioeconomic background. (Fig 4) He had yellowish keratinized plaques over the skin of his palms and soles. (Fig 1) Skin was rough and scaly. Fingers were long and slender. Lesions varied in colour and texture and underwent encrustation or fissuring.

Extra oral examination revealed normal hair and nails and a tapering face form. He had minor left sided under development of the greater alar cartilage.

Vertical dimension of occlusion was slightly reduced as was evident by the deepened mento labial fold. In addition, angular chelitis was evident on the corners of the mouth. Facial profile was retrognathic. (Fig 4) Intraoral presentation included heavy plaque deposits, halitosis, bleeding gums, pus exudates from pockets, grade 3 mobility and resorbed residual ridges. Presence of unerupted third molars was evident from bulbous tuberosities and was confirmed on radiograph.

Radiographic features include generalized severe bone loss. (Fig 2) Teeth had characteristic, "Floating in air appearance". All the remaining teeth had no bone support. The patient had unerupted third molars present in all four quadrants.

Differential diagnosis includes Haim Munk syndrome and Prepubertal aggressive periodontitis. Haim Munk syndrome is characterized by congenital palmoplantar keratoderma and progressive early onset periodontitis. It also exhibits arachnodactyly and deformity of nails and fingers in the hand. Nails and fingers were not deformed in the present case. In prepubertal aggressive periodontitis palmoplantar keratoderma is not present.[2]

Based on intra oral and extra oral findings as well as his radiographic features, the diagnosis of Papillon Lefevre syndrome was made on 21st August, 2015 in Institute of Dentistry, CMH Lahore Medical College. The diagnosis was confirmed by the patient's General Physician.

Treatment planning of this syndrome requires a multi-disciplinary team approach including dental surgeons and dermatologists. Topical treatment with emollients and 5% salicylic acid was initiated by the dermatologist. Oral acitretin(10mg/day) was also advised for dermatological management. After 8 weeks there was a marked reduction in keratoderma.

Final dental treatment plan was based upon patient's chief complaint, quality and quantity of supporting bone, affordability and age of the patient. Due to hopeless prognosis, host modulation therapy was not beneficial at this stage and extractions were planned for the remaining teeth. The patient was informed of the impacted third molars and was told that if eruption proceeds dentures will be remade. Dental implant supported prosthesis were not considered due to age of patient. However, this option was discussed with him and he was advised to pursue it after completion of his growth. Thus, conventional Complete Dentures were initially planned for this patient considering the age and affordability of the patient.[6,7]

Following dental extractions the alveolar mucosa was allowed to heal completely. The patient was re-evaluated after a recovery period of 3 months. Mucosa returned to normal healthy state(Fig 3) After assessment of residual ridges and palate, primary impression was made using admixed technique. The impression medium was a mixture of 3 parts by weight of medium fusing impression compound and 7 parts by weight of high fusing impression compound. Primary cast was poured using dental plaster and a special tray with spacer was fabricated over it. Secondary impression was taken using selective pressure impression technique with special tray using low fusing impression compound and zinc oxide impression paste. Master cast was poured using dental stone. Denture base plate and occlusal rims were constructed and maxillomandibular horizontal relation was recorded using tactile method and vertical with Niswonger's. The vertical dimension of occlusion was kept low and anterior and posterior tooth setup was done with narrow shallow cuspal acrylic teeth arranged in bilateral balanced occlusion due to excessive residual ridge resorption.

The complete maxillary and mandibular dentures were delivered to the patient (Fig 4) Denture provided adequate lip and cheek support and had excellent retention. Patient could eat and talk with it. The psychological and social well-being as well as aesthetic appearance of the patient was restored. Patient was satisfied.

A follow up was done at 3 monthly interval. At the end of first year, it was observed that there was minimal ridge resorption and denture retention was moderate with adequate chewing ability. Patient did not have any complaint with denture. However, after two years the complete dentures were remade to improve the retention and stability of dentures.

Since third molars were impacted to date, the case was kept under observation. Dentures will be revised in case of possible eruption of third molars. At completion of puberty, dental implants shall be planned to improve the quality of life for that individual.[8]

Fig 1: Keratoderma of the palms and plantar surfaces of feet.

DISCUSSION

A 16 year old male patient presented in Institute of Dentistry, C.M.H Lahore Medical College. Conventional complete dentures were selected as a treatment option. They are the treatment of choice for young edentulous patients. Similar treatment protocols for this syndrome have been reported.[2,3] Follow ups of these patients revealed that ridge resorption was minimal , masticatory efficiency was adequate and patients were satisfied.[2]

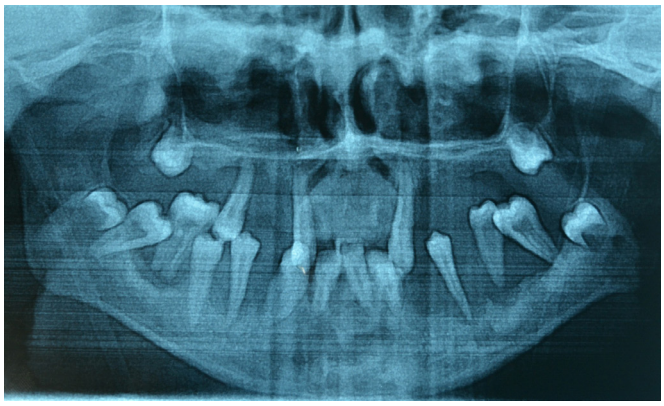

Fig 2: Orthopantomogram; Floating in air appearance of teeth.

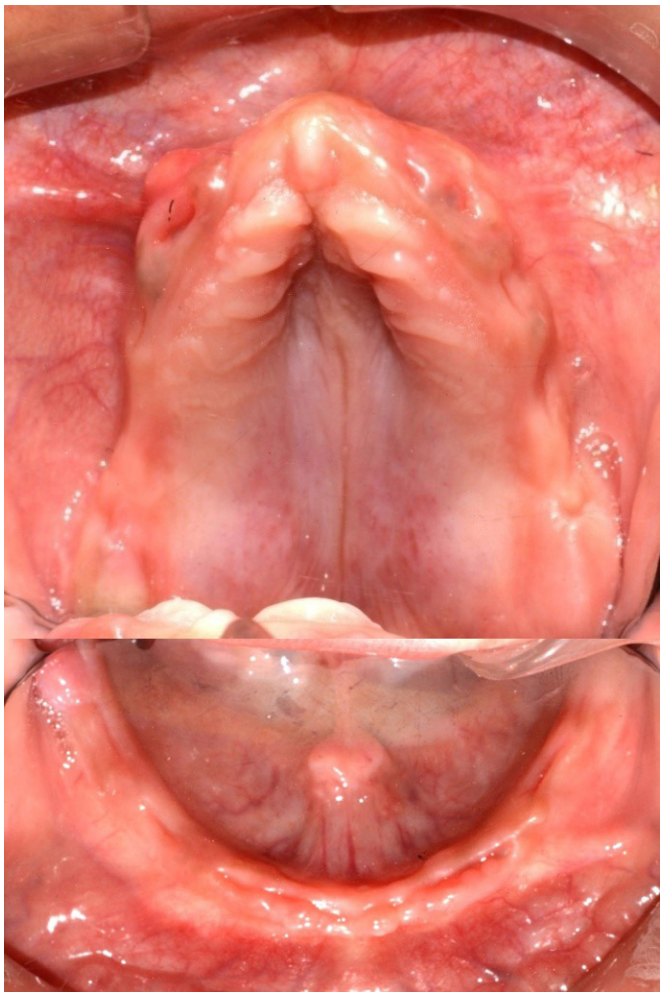

Fig 3: Intraoral view of maxillary and mandibular arch.

Conventional periodontal treatment of Papillon Lefevre syndrome has often reported to fail. However, aggressive non-surgical periodontal treatment has better success rates. This includes oral hygiene instructions, scaling and root planning and systemic anti-microbial treatment with amoxicillin, metronidazole and tetracycline. When the patient reported to

Fig 4: Frontal profile after insertion of complete maxillary and mandibular dentures.

Institute of Dentistry C.M.H Lahore, half of his teeth were already exfoliated. Remaining teeth had hopeless prognosis due to severe loss of alveolar bone. Early extractions play a role in preservation of alveolar bone if periodontitis cannot be controlled.[2,5] Thus, extractions were planned in a phased manner. This was followed by an uneventful healing period of 8 weeks.

Construction of complete denture for this patient was planned to preserve the remaining residual ridges. Admixed impression technique, shallow cusped teeth with reduced occlusal table and bilaterally balanced articulation were incorporated to improve the stability and preserve the remaining structures. Due to growth potential of individual jaws, the dentures were monitored constantly and revised.

In recent years, endosseous implants have gained popularity as important treatment alternatives. Implant placement is not contraindicated in Papillon Lefevre syndrome patients. Journal of Prosthodontics by ACP reports a 21 year old edentulous women with Papillon Lefevre syndrome who was given maxillary and mandibular fixed prosthesis supported by 16 osseointegrated implants. The results of implant treatment were successful and similar to normal individuals.8 However, growing age is a contraindication to dental implants. Implant placement is recommended after the child has reached adulthood. Otherwise, it will result in infra positioning of implants. Patient affordability is another limiting factor. Due to these reasons osseointegrated implants were not chosen as a treatment modality in this patient.

CONCLUSION

Complete Denture is a suitable prosthesis for Papillon Lefevre syndrome. It may need constant revisions depending on the stage of the disease and age of the patient. Osseointegrated implants are attractive options for the future once the patient has reached adulthood.

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CONTRIBUTIONS BY AUTHORS
All authors contributed substantialy.