Papillon-Lefevre: syndrome: Reconstruction of severely atrophic maxilla and mandible using iliac bone by Interpositional & Onlay bone graft

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Abstract

Objective: Papillon-Lefèvre syndrome is a rare autosomal recessive genetic disorder characterized by palmar-plantar hyperkeratosis, with rapidly progressive periodontitis and premature loss of both deciduous and permanent teeth. This article presents the reconstruction of jaws of a patient with Papillon-Lefèvre syndrome with iliac bone graft.

Case Report: Interpositional graft with Lefort I surgery in maxilla and Onlay bone graft in mandible in a 20-year-old female patient with Papillon-Lefèvre syndrome was performed. In our follow-up the bone grafts were clinically stable and no pain or infection was found. The patient continues to be seen at regular follow-up visits.

Conclusion: Lefort I procedure and interpositional bone graft for reconstruction of maxillary in Papillon-Lefevre syndrome, make better profile in patients. We did not only successfully treated our patient functionally and esthetically in the first stage of the treatment plan via bone grafting, but also provided psychological benefits to the patient.

Key words: Papillon-Lefèvre syndrome, Interpositional bone graft, Onlay bone graft, Lefort I.

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Introduction:

Papillon-Lefevre syndrome is a rare autosomal recessive genetic disorder accompanied by Cathepsin C gene deficiency; characterized by palmar-plantar hyperkeratosis, periodontal involvement and early teeth loss (1). In 1924, this syndrome was first described by two French physicians named Papillon and Lefevre. Its prevalence is 1-4 per million, and has no gender preference (1, 2).

This disorder is manifested by diffused palmar-plantar keratoderma and premature loss of both deciduous and permanent teeth (1, 3). Palmar and plantar keratoderma starts between the age of one to four. Erythematous keratosis plaques could appear locally, but involvement of the entire hand palm and foot sole will usually provoke bad odor. Pseudo-psoriasis plaques are also seen on knees and elbows. This condition deteriorates during winter, causing painful fissures (4). Plantar keratosis diffuses to plantar border and can also contaminate Achilles tendon’s skin. Other regions which can be involved are eyelids, cheeks, labial commissures, thighs, legs and armpits (4, 5). Usually hairs are normal in these patients, but in advanced cases, nails have transversal fissures (1, 4, 5).

Severe periodontitis is another manifestation of this syndrome appearing around the age of 3 or 4. Deciduous teeth appearance happens normally, but severe gingivitis appears without local etiology. Alveolar bone resorption begins with the second deciduous molar eruption and at the age of 4-5 all deciduous teeth are lost. This process repeats when permanent dentition appears, that’s why between the age of 10-18 the patient loses all his permanent teeth, beside the third molars, which leads to maxillary and mandibular atrophy. This article presents the reconstruction of jaws of a patient with Papilloma-Lefevre syndrome with iliac bone (7). In the past, teeth extraction was the only known
treatment for this syndrome. Because the third molars are not involved in this condition, their prophylactic extraction is not recommended and their preservation will be helpful for the patient future prosthetic treatment. Most of these patients become edentulous (except for the third molars) and will have atrophied maxilla and mandible, which causes complication in retention and stability for prosthetic treatment. For this reason, jaws’ reconstruction and bone graft have been considered in order to make implant treatment possible (7, 9, 11).

In this article report of a rare case of Papillon-Lefevre syndrome with the clinical manifestation and the bone graft treatment performed in order to use dental implants as well as a review of articles are presented.

**Case Report:**

A young 20-year-old female patient with main complaint of edentulism and primary diagnosis of Papillon-Lefevre syndrome has been referred by Shahid Beheshti Dental School department of Prosthodontics to the Oro-facial surgical department of Taleghani Hospital, Tehran, Iran. The patient dental history revealed early loss of deciduous dentition at 5 years old, followed by early loosening and loss of permanent dentition at the age of 14. The patient had no systemic particular disease and no history of previous hospitalization and surgery. In the general clinical examination of the patient, palmar and plantar hyperkeratosis has been noticed (Figures 1, 2, 3).

During oral examination, the patient was found totally edentulous, beside the presence of the right maxillary wisdom tooth, and she presented severe maxillary and mandibular ridge resorption (Figure 4).
Complete edentulism caused reduction of the longitudinal height and provoked an aging appearance (Premature senescence in appearance). In the profile view, decreased nasolabial angle and a dropped nose tip were observed (Figures 5, 6).

In the panoramic view, severe maxillary and mandibular resorption along with bilateral pneumatization of maxillary sinuses were seen (Figure 7). The Cone-beam computed tomography (CBCT) of both jaws was taken, and a common surgico-prosthetic treatment plan based on Onlay bone graft on mandible and interpositional technic on maxilla followed by fixed prosthetic treatment of both jaws was decided. In order to create and maintain suitable vertical height by the prosthodontics department, diagnostic dentures were made for both jaws.

Nasal intubation has been done under general anesthesia. Once the mouth prepared, vestibular incision on maxilla followed by muco periosteal flap has been practiced. On each side open sinus-lift surgery and then Lefort I osteotomy has been done. Maxillary bone has been relocated and protruded for 5 mm and 8 mm augmented downward in height and fixed in this appropriate position by two L-form plates on each side. A mono-cortical bone bloc measuring 3×6 cm was taken from patient’s right iliac bone and put and then fixed in the osteotomy site on the maxilla. Xeno-graft bone powder (Bio-oss, product of Geistlich Company, Suisse) was put on the grafted area; and in order to reduce resorption and to practice GBR, resorbable membrane (Bio-Gide, product of Geistlich Company, Suisse) has been used (Figure 8).

At habitual mandibular second premolar and first molar sites, in both sides, Onlay cortical bone graft has been put and fixed by screws.
Continued vertical matrix sutures have been made on the area. Presently, 5 months post-operative follow up has been performed, no graft site opening has been seen and in the radiographic evaluation the grafted bone was in suitable condition. The patient without any particular problem is a candidate for implant treatment (Figure 9).

Figure 9- Profile view after surgery

Discussion:

Papillon-Lefevre syndrome is characterized by palmar-plantar hyperkeratosis and periodontitis in childhood following eruption deciduous dentition (1-3, 8). Its pathogenesis is somehow complicated. It has been recommended to assess immunologic, genetic and microbiologic factors in the onset and evolution of this syndrome. The Cathepsin C gene mutation and its loss of function constitute effective genetic factors in occurrence of this syndrome. The Cathepsin C gene is expressed in epithelial regions such as palmar, plantar, elbow, knee and keratinized gingiva of Papillon-Lefevre syndrome which assessed (14). It is also expressed in immune cells like polymorph-nuclear and macrophages. These findings were present in the previous acute periodontitis and hyperkeratosis in the reported patient’s case. Immunologic factors cause changes in the host defense and decreased function of lymphocytes, polymorph-nuclear and monocytes’ leukocytes (2, 17, and 20).

Because in this syndrome different regions are affected, multiple services will be implicated in the treatment of these patients. Also, the early diagnosis of this syndrome has a crucial role in improving the patient’s oro-buccal health. In recent years, in patients whom dentition has not been loosed yet, various treatment plans have been introduced, including reduction of periodontal pathogenic flora by extraction of all deciduous dentition several month prior to permanent teeth eruption concurrently with an antibiotics therapy. However, unfortunately these patients are diagnosed once they lost all their permanent teeth (9). Moreover, the prosthetic reconstruction of these patients, in order to restore esthetic and nutrition function, is really important. For years, physicians have been dealing with complications in the fabrication of suitable prosthetics for atrophied maxilla and mandible in these cases, and thus tremendous efforts have been deployed to obtain new treatment plans. In addition, patients’ satisfaction especially for the lower jaw denture was poor (2). The patient referred to the Oro-facial surgery department, because she had problems with her previous dentures for both jaws wanted to have fixed prosthesis with maxillo-mandibular bone reconstruction, for esthetic purpose. After she has been informed about the eventual risk factors of such treatment, including infection, bone graft rejection and also implant failure, the patient agreed the treatment plan. The remaining upper and lower jaws both presented severe resorption, particularly in the posterior areas; which justifies iliac bone graft with interpositional technic and Lefort I surgery to maximize the skeletal esthetic and minimize graft resorption, as well as the posterior Onlay graft on lower jaw. The fixed prosthetic treatment based on dental implants is extremely efficient, regarding the esthetic, patient’s nutrition quality and her psycho-social situation (2).

Ullbro et al. (10) and also Woo et al. (11) have studied the use of dental implants in Papillon-Lefevre syndrome’s patients. Their follow-ups indicate that dental implant treatment in these patients has been successful. In another research done by Etos et al. (12), using short implants instead of bone graft, 3 years follow-up have also been reported successful.

Until now, a few researches have been published about the use of bone graft and dental implants in Papillon-Lefevre syndrome’s patients. Nevertheless, our patient is one of the rare cases
mentioned in Iran. Since the publication of this article, which was the first step of the treatment including bone graft, emotionally, the patient feels much better, and esthetically, as the nasolabial angle has been increased and para-nasal region has been augmented in volume, she feels happier. At the moment, her implant treatment plan is also ready, making her a candidate for the second step of the treatment: receiving 8 implants for the upper jaw and 8 for the lower one.

Conclusion:

As a conclusion, it could be said that based on previous studies and our patient’s case, performing bone graft in Papillon-Lefevre syndrome’s patients, in order to restore esthetic and dentition is achievable. Thus performing bone graft and dental implant treatment in these patients is recommended to other colleagues.

References:


