Prosthetic Complications in a Patient with Papillon-Lefevre Syndrome Treated with Dental Implants: A Case Report

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Introduction: Patients with Papillon-Lefevre syndrome (PLS) lose their teeth because of periodontal disease followed by alveolar bone resorption. On the other hand, this complicate implant treatment, force the surgeon to insert implants more palatally, or to do more extensive surgical procedure. **Case report:** A 24-year-old female diagnosed with PLS received an implant supported metal-acrylic prosthesis which was failed due to the dissatisfactory design and unpleasant influence on the patient function and esthetics. The prosthesis was substituted by a new designed one, fabricated by CAD/CAM technology to compensate the implants positions and fulfill patient function and esthetics. **Results**: The patient followed up the day after delivery, one week, and each 6-month, without any reported prosthetic complications or bone loss after three-year follow-up appointment. **Conclusion:** We presented the ability to restore esthetics and functions of a patient suffering from severe bone loss due to PLS by using the bone grafts and dental implants.

Keywords: Dental Implants; Full Mouth Rehabilitation; Papillon-Lefèvre Syndrome; Periodontal Treatment

Introduction

Papillon-Lefevre syndrome (PLS) is an autosomal recessive disorder which inherit by one mutated gene from each parent (1). The etiology and pathogenesis of PLS are not totally understood, but the researchers confirm that PLS is associated with cathepsin-C gene mutations, located on chromosome 11q14 (2). This rare disorder was first described by two French physicians, Papillon and Lefèvre, in France 1924 and can occur almost in one to four cases per million (3,4). Both genders are equally affected, with no specific racial tendency, in addition to normal life range (5). The characteristic of this syndrome is hyperkeratosis which could be localized or prevailed on the palms and soles. This clinical skin feature may be obvious at birth or 1 to 2 months after birth, (6) but generally appear between 6 months and 4 years of age (7). Oral manifestations commonly present when palmar-plantar hyperkeratosis exists. Such manifestations could present by rapidly progressive periodontal disease, resulting in precocious exfoliation of primary and permanent teeth which erupt in the same age and sequence as normal people with normal shape and structure. Nevertheless, PLS patients are most probably edentulous due to periodontal disease which cause hypermobility, migration, and drifting of the dentition. Eventually, PLS patients seek a prosthetic treatment in their teens (8). Newman *et al.*, found that the oral bacterial flora in these patients are gram-negative cocci, rods, and spirochetes, in addition to those found in adult periodontitis (9). Conventional periodontitis treatment, including oral hygiene instructions, scaling and root planning, and periodontal surgery, and systemic antibiotics usually fail in individuals with PLS. On the other hand, severe alveolar bone resorption will be the result of the unrestricted progress of the periodontal disease (10). For this reason, some clinicians recommend to have early extractions of all permanent teeth to save the remaining bone in order to facilitate the implant treatments (11). Simple prosthetic treatment options for PLS patients could be a combination of conventional complete dentures and overdentures. In recent years, oral rehabilitation by placement of endosseous dental implants have taken a major

place in treatment options of edentulous jaws for their great benefit in providing support, stability, and retention of prostheses. Using dental implants in PLS affected patients have been reported in few cases, and the results of this type of treatment demonstrated that implants could be indicated as a successful option for PLS patients (11-16).

Furthermore, good prognosis of dental implant treatments has been shown in patients with PLS and severe periodontitis (14). An important factor for success of implant treatment is available bone; which in these patients usually have severe atrophy (17). Therefore, according to the previous reported cases (11,18) this led to treatment with implant-supported overdenture in order to overcome the increased restoration space due to severe bone resorption. The overdenture prosthesis could facilitate the treatment procedures and decrease the cost and the duration of the treatment compared to fixed option. Moreover, choosing a fixed implant-supported prosthesis as the treatment option in these patients, put us in more challenging situation because of complicated procedures, such as bone augmentation, nerve repositioning in the posterior mandible, using short implants, and sinus lifting in the posterior maxilla (19-21).

The aim of this study was to detail the prosthetic complication management of a young edentulous PLS patient, previously treated with implant supported maxillary and mandibular fixed dental prosthesis.

Case Report

A 24-year-old female diagnosed with PLS was referred to the prosthodontics department, Tehran University of Medical Science, Tehran, Iran. The patient was treated with 8 implants in the maxilla and 6 implants in the mandible (Ihde, Dr Ihde Dental GmbH, Eching, Germany) which supported a metal-resin fixed (hybrid) prosthesis for one year (Figure 1). The patient's chief complaint was "I don't like my current prosthesis, it is big, ugly and I have difficulty in eating and speaking". She had hyperkeratosis on her palms and soles regions without any other remarkable medical history or abnormal findings. She lost her teeth due to progressive periodontal disease. The patient had received augmentation surgery of mandibular bone simultaneously with implants insertion. She had received fixed metal-resin prosthesis, though the result was not meeting her expectations esthetically and functionally. Furthermore, the patient was not fully comfortable with speaking due to bulky palatal part of her maxillary prosthesis (Figure 2).

Figure 1. Frontal view of the metal-resin prosthesis

All these reasons prompted her to substitute her old prosthesis with new one, ensured the esthetic and function for her.

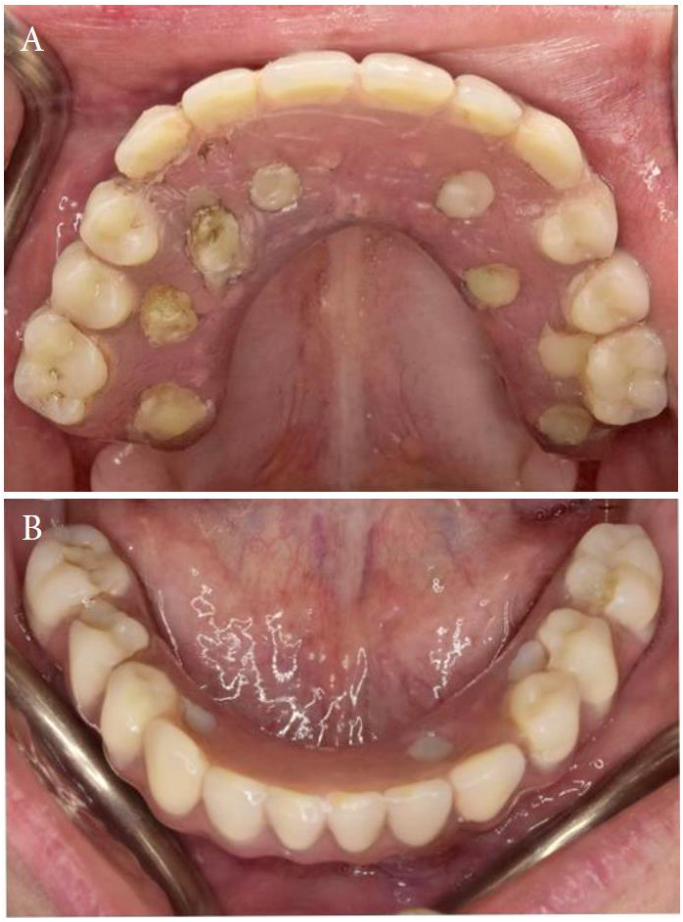

Figure 2. (A) Occlusal view of the metal-resin prosthesis (maxilla); (B) Occlusal view of metal-resin prosthesis (mandible)

Intraoral examination disclosed bulky fixed prosthesis, multi repair of fractured teeth, and gingival enlargement around the anterior left and posterior right implants. After removing the lower prosthesis, the enlarged gingiva was removed under local anesthesia and healing abutments screwed to give the soft tissue chance to heal. Several attempts to remove the upper prosthesis had failed due to striping in one of the screw heads which left the prosthesis stuck in her mouth. Implant repair and help kit (22), ultrasonic tip with water irrigation (23) were used to lose the screw but there was no success; so, a vertical groove was cut on the screw head in order to use flat blade screwdriver (24) though this attempt did not also work as it was not possible to have a good access to the screw which was so deep. Finally, the prosthesis was cut into two parts, the abutment was prepared to expose the screw, and the screw was unfastened with hemostatic forceps and the abutment with the screw were removed (Figure 3).

Figure 3. Screw head stripping

After removing the stuck prostheses, primary impressions with alginate (Zhermack,Badia Polesine, Italy) for implant-supported prostheses were made. Custom open trays (SR Ivolen, Ivoclar, Vivadent, Schaan, Liechtenstein) were fabricated on the primary casts for final impressions. Open tray impression copings (Ihde, Dr Ihde Dental GmbH, Eching, Germany) were fastened intraorally, and final impressions were made with polyvinyl siloxane (Panasil® initial contact Light, Kettenbach, Germany) after splinting the impression copings together using auto-polymerizing resin (GC Pattern Resin; GC America Inc., Alsip, IL). The impressions were poured with type IV dental stone (Prima-Rock, Whip Mix Corp., Louisville, KY), then verification jigs were made to check the accuracy of the impressions. The master casts were mounted on the articulator using the face-bow and screw-retained record bases for centric relation.

Try in session were held for checking the denture teeth arrangement after removing the record base flanges to confirm the ability of fix restoration as the treatment plan. The final cast were scanned (Dentsply Sirona, inLab scanner, Bensheim, Germany); then PMMA (Dentsply Sirona, Bensheim, Germany) screw retained temporary restoration were designed (Exocad GmbH, DentalCAD,Darmstadt, Germany) and milled (Arum 5x 200, Arum Europe GmbH), to be as template for the final restoration in order to check the passive fit, esthetics, phonetic, occlusion, and the patient profile which will allow to reduce the needed modifications of the final restoration. After the patient and her family approved the temporary prostheses, it was sent to the lab to continue the procedure. The lab technician scanned the cast with the PMMA restoration in place and cutback (1.7mm) was done to the scan and cobalt-chrome (Co-Cr) framework (Starbond CoS disc, Scheftner dental alloys, Germany) was milled. The metal frameworks (Figure 4).

Figure 4. Metal framework try-in

Were evaluated intraorally to ensure the passivity without "rocking" movement by the alternate pressure technique and taking radiographs after applying one-screw test (25). The next step was ceramic layering, using feldspathic porcelain in B1 shade according to the manufacturer's instructions (Vita Omega Metal Ceramics, Vita Zahnfabrik, Bad Sackingen, Germany) for the teeth and soft tissue area using silicon index made from the temporary restorations in order to have the same contour.

Followed by clinical bisque bake, the prostheses were tried in to modify and confirm the mutually protected occlusion, centric relation, contour, esthetics, phonetic and the shade. The metal-ceramic prostheses were finally characterized and glazed. At delivery time, the prostheses (Figure 5) screwed into implant bodies and torqued (30 Ncm), as suggested by the manufacturer. Teflon tapes were placed over the screws access holes and covered with light-cured composite resin (Z250, 3M ESPE, Minnesota, United States). Panoramic radiograph was ordered and postoperative hygienic instructions were illustrated to the patient.

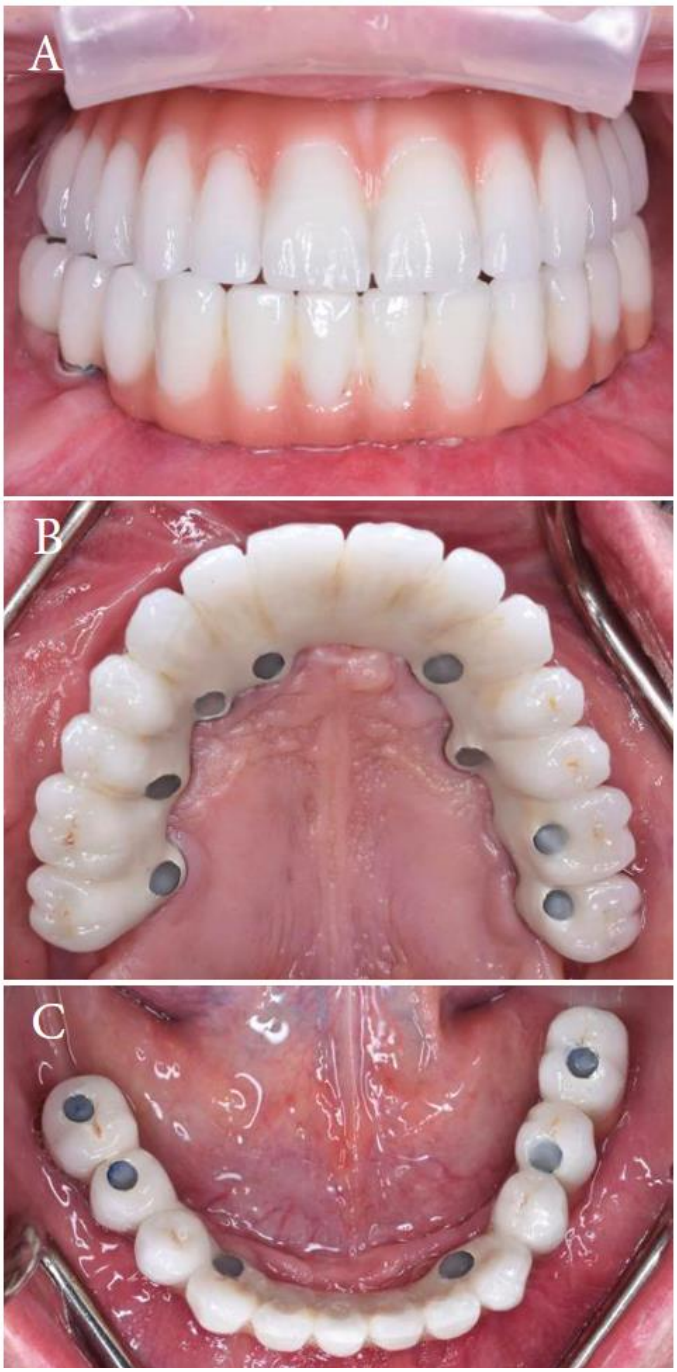

Figure 5. (A) Frontal view of final prosthesis; (B) Occlusal view of final prosthesis (maxilla); (C) Occlusal view of final prosthesis (mandible)

Results

The patient followed up the day after delivery, one week, and each 6-month, without any reported prosthetic complications or bone loss after three-year follow-up appointment.

Discussion

The individuals who affected with PLS will be psychologically, physically, and socially compromised due to the syndrome manifestations. Oral rehabilitation for this type of patients would be through conventional partial or complete denture. On the other hand, more promising treatment prognosis could be achieved through osseointegrated implants which assure more pleasant outcome especially when the patients are in their first decades of life and have increased esthetic demands like the female in this report (26). Dental implants become a routine practice with high success rate. The essential factor for success after osseointegration is the biomechanical consideration for the prosthetic aspect which compose of crown-implant ratio (CIR), crown height space (CHS), cantilevers, cusps and occlusal table shape, overload and off-axis loads. These biomechanical factors especially CHS and CIR are more critical in patients with PLS because of severe resorption in the alveolar bone. Therefore, the treatment plan will be more challenging particularly when the patient has the desire to restore function and esthetics by fixed prosthesis. Consequently, the surgeons have to do bone grafting, use shorter implants, and increase the number of implants to compensate the increased CHS (27-28). Finite element analysis studies of Verri *et al.*, and Bulaqi *et al.*, showed that increasing CHS increased the stress concentration on bone tissue and prosthetic screw especially with non-axial forces (29-30). Other studies evaluated the CIR and they concluded that CIR has no critical effect on bone loss around the implants; and short

implants have similar success rate as standard implants (28,19-21).

As implant placement is not a contraindication for PLS patients, this case had received dental implants and to overcome the biomechanical complications and reduce the stress in the peri-implants bone, the implants were splinted together (31-34). In the last decades, the application of computer-aided design/computer-aided manufacture (CAD/CAM) significantly increased in the dental field (35). CAD/CAM technology allowed the practitioners to use a wide range of new materials. Furthermore, these new materials improved our treatment plan options because it was not available for traditional techniques (36).

The use of CAD/CAM in dentistry has abundant features over traditional techniques, such as the ease of use, speed, quality and less technique sensitivity. Furthermore, the subtractive milling process of CAD/CAM materials eliminate the flaws that could happen during the process in the conventional techniques, such as pouring, wax-up, investing, firing, metal casting, and polishing (37). It has been confirmed that increasing the fabrication steps will introduce more probabilities of inaccuracies in the final restoration (38). CAD/CAM system is distinguished by decreasing these fabrication steps. Moreover, it does not rely on human work while the CAD/CAM will help the clinician in designing and fabricating the restorations (37).

In this clinical report two reasons for complications of former metal-resin prosthesis could be enumerated; first, palatal position of implants in the maxilla because of severe bone resorption and the second, increased CHS of near 15mm (from the implant platform to the occlusal plane) in each jaw. In the previous restoration, the clinician tried to overcome the increased CHS by using metal-resin prosthesis in order to decrease the size of framework to prevent any deformation during casting. CAD/CAM technology allowed us to mill frameworks in increased CHS without having any concern about metal distortion and deformation due to significant contraction and expansion related to temperature fluctuation during investing and casting procedure of large amount of metal alloy (39-40).

Furthermore, appropriate communication between the surgeons, prosthodontists, lab technician, and the patient with a proper treatment plan, and logical expectation from the patient will lead to a satisfactory outcome for all the parties and eliminate time and cost consuming by repeating the treatment. Moreover, emphasizing the importance of oral hygiene especially for PLS patient is critical for long term success of treatment.

Conclusion

In this case, we presented the ability to restore esthetics and functions of a patient suffering from severe bone loss due to PLS by using the bone grafts and dental implants. The final result exceeded the patient expectations and substantially enhanced her quality of life and had a positive reflection on her personality.

Conflict of Interest: 'None declared'.

References

1. Wani AA, Devkar N, Patole MS, Shouche YS. Description of two new cathepsin C gene mutations in patients with Papillon-Lefevre syndrome. J Periodontol. 2006;77(2):233-7.
2. Canger EM, Celenk P, Devrim I, Yenisey M, Gunhan O. Intraoral findings of Papillon-LeFevre syndrome. J Dent Child 2008;75(1):99-103
3. Kaur B. Papillon Lefevre syndrome: a case report with review. Dentistry. 2013;3(1):161-122.
4. Gorlin RJ, Sedano H, Anderson VE: The syndrome of palmar-plantar hyperkratosis and premature periodontal destruction of the teeth. J Pediatr 1964;65(6):895-898.
5. Haneke E: The Papillon-Lefevre syndrome: keratosis palmoplantaris with periodontopathy. Report of a case and review of the cases in the literature. Hum Genet 1979;51(1):1-35
6. Galanter DR, Bradford S: Case report. Hyperkeratosis palmoplantaris and periodontosis: the Papillon-Lefevre Syndrome. J Periodontol 1969;40(1):40-47
7. Wiebe CB, Häkkinen L, Putnins EE, Walsh P, Larjava HS. Successful periodontal maintenance of a case with Papillon-Lefevre syndrome: 12-year follow-up and review of the literature. J Periodontol. 2001;72(6):824-30.
8. Sreeramulu B, Shyam ND, Ajay P, Suman P. Papillon–Lefèvre syndrome: clinical presentation and management options. Clin cosmet Investig Dent. 2015;7(1):75-81.
9. Newman M, Angel I, Karge H, Weiner M, Grinenko V, Schusterman L. Bacterial studies of the Papillon-Lefevre syndrome. J Dent Res. 1977;56(5):545-545.
10. Schacher B, Baron F, Ludwig B, Valesky E, Noack B, Eickholz P. Periodontal therapy in siblings with Papillon–Lefevre syndrome and

tinea capitis: a report of two cases. J Clin Periodontol. 2006;33(11):829-36.
11. Woo I, Brunner DP, Yamashita DD, Le BT. Dental implants in a young patient with Papillon-Lefevre syndrome: A case report. Implant Dent. 2003;12(2):140-4.
12. Ahmadian L, Monzavi A, Arbabi R, Hashemi HM. Full-Mouth Rehabilitation of an Edentulous Patient with Papillon-Lefèvre Syndrome Using Dental Implants: A Clinical Report. J Prosthodont. 2011;20(8):643-8.
13. Shah J, Goel S. Papillon-Lefevre syndrome: Two case reports. Indian J Dent Res. 2007;18(4):210-213.
14. Ullbro C, Crossner CG, Lundgren T, Stålblad PÅ, Renvert S. Osseointegrated implants in a patient with Papillon-Lefèvre syndrome: A 4½-year follow-up. J Clin Periodontol: Case Report. 2000;27(12):951-4.
15. Van der Weijden GA, Van Bemmel KM, Renvert S. Implant therapy in partially edentulous periodontally compromised patients: A review. J Clin Periodontol. 2005;32(5):506-511.
16. Senel FC, Altintas NY, Bagis B, Cankaya M, Pampu AA, Satıroglu I, Senel AC. A 3-year follow-up of the rehabilitation of Papillon-Lefèvre syndrome by dental implants. J Oral Maxillofac Surg. 2012;70(1):163-7.
17. Orsini G, Bianchi AE, Vinci R, Piattelli A. Histologic evaluation of autogenous calvarial bone in maxillary onlay bone grafts: a report of 2 cases. Int J Oral Maxillofac Implants 2003;18(4):594-598.
18. Etöz OA, Ulu M, Kesim B. Treatment of patient with Papillon-Lefevre syndrome with short dental implants: a case report. Implant Dent. 2010;19(5):394-9.
19. Fan T, Li Y, Deng WW, Wu T, Zhang W. Short implants (5 to 8 mm) versus longer implants (> 8 mm) with sinus lifting in atrophic posterior maxilla: a meta-analysis of RCTs. Clin implant Dent Relat Res. 2017 Feb;19(1):207-15.
20. Tong Q, Zhang X, Yu L. Meta-analysis of Randomized Controlled Trials Comparing Clinical Outcomes Between Short Implants and Long Implants with Bone Augmentation Procedure. Int J Oral Maxillofac Implants. 2017;32(1):25-34.
21. Nisand D, Picard N, Rocchietta I. Short implants compared to implants in vertically augmented bone: a systematic review. Clin Oral Implants Res. 2015;26(1):170-9.
22. Gooty JR, Palakuru SK, Guntakalla VR, Nera M. Noninvasive method for retrieval of broken dental implant abutment screw. Contemp Clin Dent. 2014;5(2):264.
23. Walia MS, Arora S, Luthra R, Walia PK. Removal of fractured dental implant screw using a new technique: a case report. J Oral Implantol. 2012;38(6):747-50.
24. Williamson RT, Robinson FG. Retrieval technique for fractured implant screws. J Prosthet Dent. 2001;86(5):549-50.
25. Kan JY, Rungcharassaeng K, Bohsali K, Goodacre CJ, Lang BR. Clinical methods for evaluating implant framework fit. J Prosthet Dent. 1999;81(1):7-13.
26. Elsayed A, Wille S, Al-Akhali M, Kern M. Comparison of fracture strength and failure mode of different ceramic implant abutments. J Prosthet Dent. 2017;117(4):499-506.
27. Jokstad A, Braegger U, Brunski JB, Carr AB, Naert I, Wennerberg A. Quality of dental implants. Int Dent J. 2003;53(6):409-43.
28. Nissan J, Ghelfan O, Gross O, Priel I, Gross M, Chaushu G. The effect of crown/implant ratio and crown height space on stress distribution in unsplinted implant supporting restorations. J Oral Maxillofac Surg. 2011;69(7):1934-9.
29. Verri FR, Junior JF, de Faria Almeida DA, de Oliveira GB, de Souza Batista VE, Honório HM, Noritomi PY, Pellizzer EP. Biomechanical influence of crown-to-implant ratio on stress distribution over internal hexagon short implant: 3-D finite element analysis with statistical test. J Biomech. 2015;48(1):138-45.
30. Bulaqi HA, Mashhadi MM, Safari H, Samandari MM, Geramipanah F. Effect of increased crown height on stress distribution in short dental implant components and their surrounding bone: A finite element analysis. J Prosthet Dent. 2015;113(6):548-57.
31. Nissan J, Gross O, Ghelfan O, Priel I, Gross M, Chaushu G. The effect of splinting implant-supported restorations on stress distribution of different crown-implant ratios and crown height spaces. J Oral Maxillofac Surg. 2011;69(12):2990-4.
32. Bal BT, Çağlar A, Aydın C, Yılmaz H, Bankoğlu M, Eser A. Finite element analysis of stress distribution with splinted and nonsplinted maxillary anterior fixed prostheses supported by zirconia or titanium implants. Int J Oral Maxillofac Implants. 2013;28(1):27-38.
33. Lee JB, Kim MY, Kim CS, Kim YT. The prognosis of splinted restoration of the most-distal implants in the posterior region. J Adv Prosthodont. 2016;8(6):494-503.
34. Behnaz E, Ramin M, Abbasi S, Pouya MA, Mahmood F. The effect of implant angulation and splinting on stress distribution in implant body and supporting bone: A finite element analysis. Eur J Dent. 2015;9(3):311-318.
35. Davidowitz G, Kotick PG. The use of CAD/CAM in dentistry. Dent Clin North Am. 2011;55(3):559-70.
36. Wittneben JG, Wright RF, Weber HP, Gallucci GO. A systematic review of the clinical performance of CAD/CAM single-tooth restorations. Int J Prosthodont 2009; 22(5):466–71 [LSEP]
37. Abduo J. Fit of CAD/CAM implant frameworks: a comprehensive review. J Oral Implantol. 2014;40(6):758-66.
38. Michalakis KX, Hirayama H, Garefis PD. Cement-retained versus screw-retained implant restorations: a critical review. Int J Oral Maxillofac Implants. 2003;18(5):719–728.
39. Drago C, Saldarriaga RL, Domagala D, Almasri R. Volumetric determination of the amount of misfit in CAD/CAM and cast implant frameworks: a multicenter laboratory study. Int J Oral Maxillofac Implants. 2010;25(5):920–929.
40. Torsello F, di Torresanto VM, Ercoli C, Cordaro L. Evaluation of the marginal precision of one-piece complete arch titanium frameworks fabricated using five different methods for implant-supported restorations. Clin Oral Implants Res. 2008;19(8):772–779.

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