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Case Report

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Prosthetic rehabilitation with dental implant and fixed prosthesis for Apert syndrome patient: A case report

Abdulwahab I Al-kholani[1]; Soumia A Al-Maghreby[2]; Mohja A Monibari[3]; Taha A Alansi[4]; Mohamed Al-hemiari[5]; Seham M Shamlan[3]

[1]Professor of Restorative Dentistry, Dental Implant Consultant, Faculty of Dentistry, Sana'a University, Sana'a, Yemen.

[2]Professor, Abdulwahab Al-kholani Dental Center, Sana'a, Yemen.

[3]Private Dental Laboratory, Sana'a, Yemen.

***Corresponding Author: Abdulwahab Ismail Al-kholani**

Faculty of Dentistry, Conservative Department, Sana'a University, Sana'a, Yemen

Email: prof_kholani@yahoo.com

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Abstract

Apert syndrome is characterized by the premature fusion of certain skull bones (craniosynostosis), cone shaped calvarium, mid-face hypoplasia, ocular manifestations and symmetric syndactyly of hands and feet. This early fusion prevents the skull from growing normally and affects the shape of the head and face with class III malocclusion. In addition, a varied number of fingers and toes are fused together (syndactyly). A prosthetic management of the condition is arguably more economical than complex surgical treatments when the patient's demand is more on esthetic correction, especially in the advanced stage. In this report, we describe a case involving a 37-year-old woman who showed good treatment outcomes after full mouth rehabilitation including endodontic, dental implant and fixed prosthodontic treatment to overcome the problem of maxillary hypoplasia, severe maxillary arch constriction and severe occlusal collapse.

Keywords: Apert syndrome; craniosynostosis; syndactyly; fixed prosthesis; dental implant.

Introduction

Apert syndrome is named for the French physician. Eugene Apert in 1906 described the syndrome acrocephalosyndactyly [1]. Apert syndrome most frequently associated with craniosynostoses. The term craniosynostosis refers to early closure of cranial sutures. The cranial malformations are the most apparent effects of acrocephalosyndactyly. Patients with this syndrome present severe syndactyly, exophthalmia, ocular hypertelorism and hypoplastic midface with Class III malocclusion and systemic alterations [2,3]. In addition genetic factor play a role in this syndrome [4].

Case report

A 37-year-old woman presented with complaints of symmetric syndactyly of both hands and feet, abnormal head shape, exophthalmia, ocular hypertelorism and severe class III malocclusion. Clinical, extra oral, and intraoral examination revealed all abnormal features which where characteristic of Apert syndrome. In addition to that patient had missing teeth in relation to maxillary and mandibular right and left posterior regions which were extracted several years back (Figure 5).

No family history of similar complaints or any other hereditary abnormality was reported. Patient complaining of difficulty in chewing food, mal-aligned teeth and facial deformity. Detailed history revealed that she was the first child of an apparently normal mother. No history of consanguineous marriage of her parents was reported. Her medical history revealed that she has been suffering from these problems since birth and had never undergone any corrective surgery intra orally. Corrective surgery was performed for the fingers only in both hands.

Examination revealed Ocular proptosis, strabism, hyper-

telorism, incompetent everted lips and bilateral symmetrical syndactyly were present (Figure 2). In addition intraoral examination showed severe class III malocclusion with reverse overjet (Figure 1) and bulky high arched V-shaped palate with submucosal occult cleft.

Soft tissue alterations were observed which represented the typical lateral palatal swellings (Figure 3). There was no cleft palate.

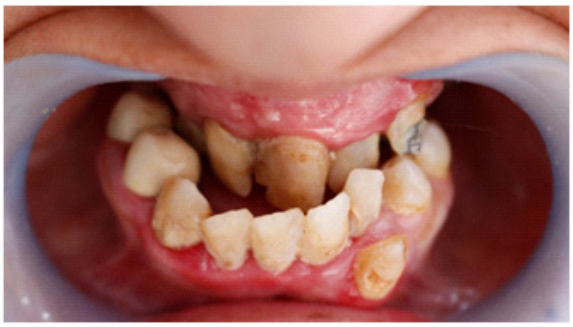

Figure 1: Intermaxillary relationship before treatment.

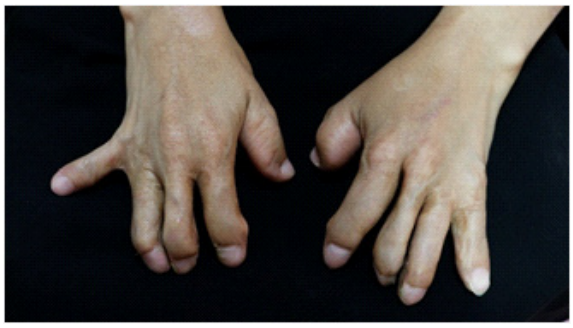

Figure 2: Bilateral symmetrical syndactyly.

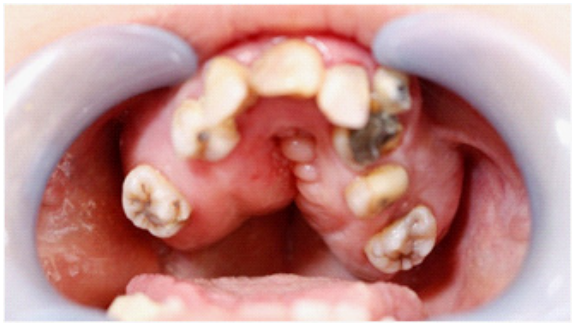

Figure 3: Maxillary arch before treatment with lateral palatal swellings.

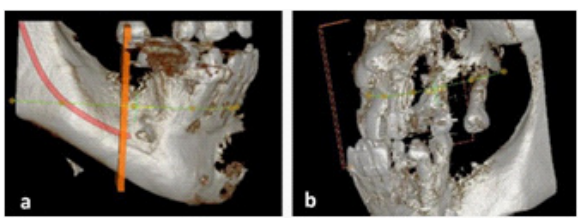

Figure 4: CBCT radiograph

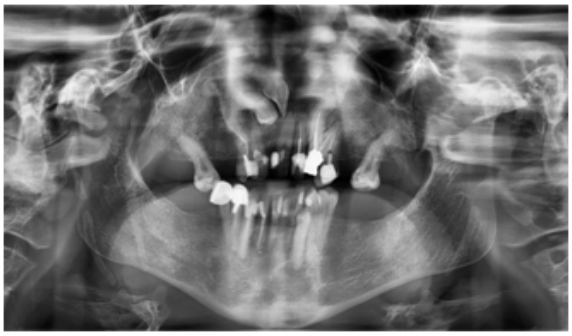

Figure 5: Panoramic radiograph.

Procedure

A prosthetic management of the condition is arguably more economical than complex surgical treatments when the patient's demand is more on esthetic correction, especially in the advanced stage.

On investigation, Cone Beam Computed Tomography (CBCT) (Figure 4a and b) and panoramic radiograph (Figure 5) revealed maxillary deformity with malaligned teeth.

Patient's parents agreed with the treatment plan and informed consent were obtained from them. Accordingly the treatment was initiated.

Maxillary and mandibular impressions were made with alginate impression material. Casts were mounted on an articulator. Implant presurgical diagnostic procedures were completed, and it was decided to place 2 implants in each mandibular sites.

Single piece immediate loading implants were used. The first implant in second premolar region and the second implant in first molar region in both mandibular sides (Figures 6).

After implantation, teeth were prepared for the purpose of full mouth rehabilitation. All teeth were prepared with chamfer type margins. Two types of impressions were made an alginate impression for an acrylic transitional prosthesis and silicone impressions for final Porcelain Fused to Metal (PFM) restoration.

Among the malpositioned and severely tipped teeth, endodontic treatment was performed for proper tooth preparation.

The try in for metal framework for mandibular and maxillary full arches restorations were done as in routine. Maxillary wax rims were prepared over the retentive framework in the usual manner. The height of the rims, lip support, and inclination of the occlusal plane were adjusted in the mouth before determining occlusal vertical dimension. The occlusal vertical dimension was then determined. The centric relation was recorded with interocclusal registration method.

The maxillary and mandibular fixed prosthesis was finished and glazed with conventional procedures followed by final cementation (Figures 7, 8 and 9).

The patient was successively reviewed post-treatment in a sequence of one day, one week and one month. Reviews evidently showed a psychological improvement in the patient. Patient was visibly socializing better and also initiated interactions with others.

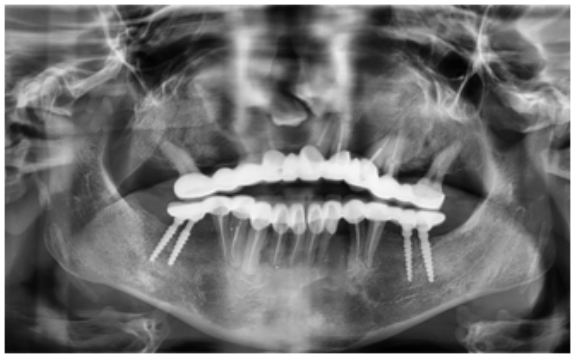

Figure 6: Single piece implants in mandibular right and left regions.

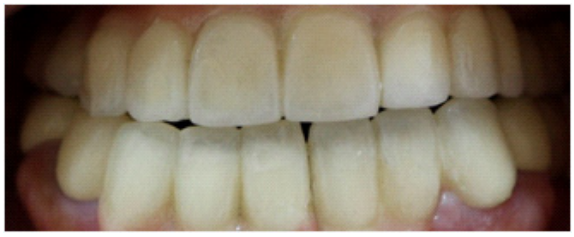

Figure 7: Single piece implants in mandibular right and left regions.

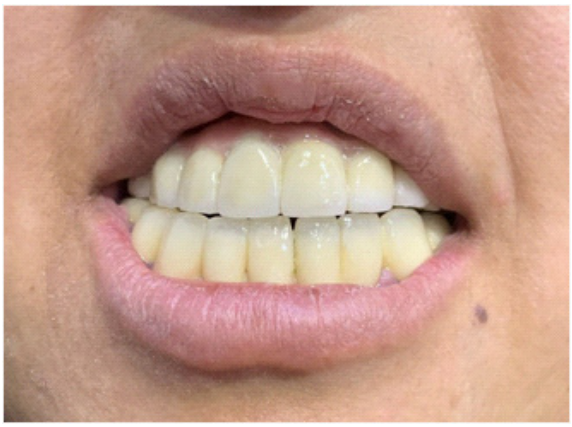

Figure 8: Post treatment frontal view.

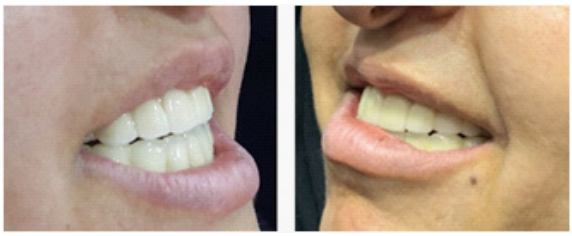

Figure 9: Post treatment frontal view.

Discussion & conclusion

Treatment planning for Apert patients varies according to the age the syndrome is diagnosed. The treatment of Apert syndrome must commence at birth or at the least in the pediatric stages and a life-long management supervised by a team of healthcare specialists must be ensured [5]. The challenges significant to conventional dentistry is the complex relationship between the supporting basal maxillary and mandibular bones as well as the pseudo or real cleft palate.

The dental management of this syndrome is marked by its unique and integrated treatment plans because each patient manifests unique physical and mental challenges that requires an individualized, comprehensive and collaborative care.

The present case exemplifies the centrality of individualized and tailored treatment plans for Apert syndrome. The patient was neither administered any treatment at birth nor been consulted for diagnosis throughout her childhood. Lack of long-term intensive care has resulted in the patient's loss of dentition. Therefore, orthodontic management falls counterproductive. Missing teeth on the other hand, opens a way for prosthodontic rehabilitation.

There were certainly more complicated treatment options, which mostly included quiet invasive orthognatic surgical procedures [6]. As the patient refused surgical treatment, these options were initially eliminated. However, in adult patients surgical management may not be preferred for various reasons by the patient, as in our case.

Usually, multiple surgeries to prevent maxillary hypoplasia and cranial synostosis are planned starting from the early ages. But, for the adolescent and adult patients, little is achieved with surgery [7].

Prosthodontic treatment considered to be a good choice to restore not only the dental arches but also facial contours as well for selected patients with Apert Syndrome. The type of prostheses, the supporting structures or implants should be evaluated for each patient.

In case of Apert Syndrome the utilization of existing teeth for retention, stability, function and the phonetics should be considered. In addition, in this case, dental implant was considered at the sites of missing teeth.

Prosthetic reconstruction of function and aesthetics for Apert patients avoiding further surgical applications appears out to be a powerful alternative to improve the quality of life and patients' satisfaction.

This case study also demonstrates the psychosomatic implications of a successful prosthetic rehabilitation of Apert syndrome as the patient exhibited visible behavioral progress post reclamation of the aesthetics of her face [8].

Declarations

Patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form the patient have given her consent for her images and other clinical information to be reported in the journal. The patients understand that her name and initial will not be published.

Conflict of interest: No potential conflict of interest is relevant to this article, all authors declare that no financial and personal relationships with other people or organizations could inappropriately influence (bias) their work. Examples of potential conflicts of interest include employment, consultancies, stock ownership, honoraria, paid expert testimony, patent applications/registrations, and grants or other funding.

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