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RESEARCH ARTICLE

PROSTHETIC TREATMENT OF A PATIENT WITH NAGERACROFACIALDYSOSTOSIS: A CASE REPORT

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Abstract

Introduction: Nager syndrome, also known as Nager acrofacialdysostosis, was first described by Nager and de Reynier in 1948. It is a rare syndrome resulting from developmental abnormalities of the first and second branchial arches and is mostly sporadic. The prevalence is unknown; about 100 cases have been published up to now. The purpose of this report is to present a case of Nager syndrome where we were able to achieve a conventional complete denture despite the many challenges due to the complexity of the clinical case.

Case Report: A 27-years-old female patient was referred to our service of Removable Prosthodontics at Casablanca University Hospital Center complaining of difficulty in speaking and chewing. The patient presented several craniofacial anomalies including facial atrophy, maxillomandibular hypoplasia, flat nasal bridge, associated to defects upper and lower limbs. Based on these craniofacial characteristics and the coexisting upper and lower limb preaxial anomalies, a diagnosis of Nager syndrome was confirmed. The intra-oral examination revealed an edentulous mandibular arch and carious, mobile and hypomineralized maxillary teeth, the patient presents a severe micrognathia with a skeletal CI III which complicates the prosthetic management of the patient. The realization of a complete denture has improved the comfort and function of the patient.

Conclusion: Nager Syndrome is a condition with a rather heavy clinical situation and its management must be multidisciplinary, the psychology and comfort of these patients is often affected. Oral care has improved aesthetics, function and the quality of life in this patient with Nager syndrome.

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Introduction:-

Nager syndrome, also known as Nager acrofacialdysostosis (NAD), was first described by Nager and de Reynier in 1948 [11]. It is a rare syndrome resulting from developmental abnormalities of the first and second branchial arches and is mostly sporadic; however, autosomal dominant or autosomal recessive inheritance has been reported. Nager syndrome has an alteration of the 9q32 chromosome, 1q12q21 deletion. The prevalence is unknown; about 100

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cases of Nager syndrome have been published up to now. Patients with Nager syndrome are found worldwide among all racial and ethnic groups [2].

The main clinical features consist of craniofacial, limb, and musculoskeletal anomalies. Less frequently, other malformations have also been reported.

Craniofacial anomalies:

The facial features of NAD are distinctive; cardinal features include downward slanting palpebral fissures, malar hypoplasia, a high nasal bridge, micrognathia, and external ear defects.

Extension of scalp hair onto the cheeks, a reduced number of eyelashes and lower lid colobomas occur less frequently. The most common external ear anomalies include external auditory canal stenosis and low set positioning. Posterior rotation and preauricular skin tags have been observed less frequently [3,5,7].

Hearing loss is an important feature; it is typically bilateral, conductive, and of the order of 50 to 70 dB. Even in the absence of external ear anomalies, ossicular chain malformations may occur.

The predominant oral findings include cleft palate and an absent soft palate. Cleft lip, velopharyngeal insufficiency, and foreshortening of the soft palate occur less frequently. Hypoplasia of the larynx or epiglottis occurs rarely, Temporomandibular joint fibrosis and ankylosis may occur [6].

In addition to hypoplasia, the maxilla is posteriorly malrotated and often associated with protrusion of the central upper midface [9].

Limb defects:

Limb anomalies are a cardinal sign of NAD and, in combination with the characteristic facial features, are diagnostic. Typical limb abnormalities include preaxial anomalies (hypoplastic or absent thumbs and radii) and proximal radioulnar synostosis. Thumb anomalies are usually asymmetrical. Duplicated and triphalangeal thumbs may occur [10].

Case Report

A 27-years-old female patient was referred to the service of Removable Prosthodontics in Casablanca University Hospital Center with complaint of dental caries and difficulty in speaking and chewing.

The patient presented several craniofacial anomalies including facial atrophy, maxillomandibular hypoplasia, severe micrognathia, malar hypoplasia, flat nasal bridge, cleft palate (Fig1).

No family history regarding any craniofacial disorder neither consanguinity has been reported.

The patient presented other features including respiration, feeding, and speech problems as well as short fore arms and deformity of the hands and feet with some fingers absent and some fused together (syndactyly) (Fig2).

There was no mental retardation. Intraoral examination revealed a major problem of patient's oral hygiene. She had multiple carious maxillary teeth with mobility and hypomineralisation. All mandibular teeth were extracted after a carious process, we noted restricted mouth opening, severe micrognathia with skeletal class III and space deficiency in maxillary and mandibular arch (Fig3,4).

Based on the craniofacial characteristics and the coexisting upper and lower limb preaxial anomalies, a diagnosis of Nager syndrome was confirmed.

Oral hygiene education was given to the patient and orthodontic consultation was held. The patient was instructed about severity of surgical treatment. When given the options of pre-prosthetic surgery and dental implants, patient refused to undergo a surgery and requested a non-surgical treatment option. Consequently, conventional complete denture therapy was undertaken.

The carious maxillary teeth were extracted, and firstly provisional prosthesis was made after healing tissue. Primary and secondary impressions was done trying to make maximum use of the support surface and the undercut areas in the maxilla to ensure subsequent retention of the prosthesis (Fig5).

The registration of jaw relations was delicate given the significant skeletal Class III, and therefore a compromise situation had to be found to ensure esthetics and function, the temporary prosthesis was used to guide this step of registration of jaw (Fig6,7).

Figure 1: Front and side view showing the patient's facial dysmorphism.

Figure 2:- Defects of hands and feet.

Figure 3:- Edentulous mandibular arch, carious maxillary teeth with hypomineralization.

Figure 4:- Inter-maxillary report.

Figure 5:- Maxillary and mandibular secondary impressions with Kerr paste and impression paste.

Figure 6:- Registration of jaw relations.

Figure 7:- Insertion of complete denture and patient's satisfaction.

Discussion:-

Nageracrofacialdysostosis is a rare congenital syndrome and nearly 100 cases have been reported in the medical literature. Nager syndrome is characterised by craniofacial, limb and musculoskeletal anomalies. The orofacial features are similar to those of Treacher Collins syndrome, but it can be distinguished from Treacher Collins syndrome by preaxial limb deformities.

Nageracrofacialdysostosis is associated with various syndromic deformities, making it difficult to predict the treatment result. [4]

In approximately 50% of patients, NAFD is associated with heterozygous mutations in the SF3B4 gene (1q12q.21), encoding a component of the splicing complex.

The diagnosis of NAFD is based on physical and radiological examination, or the identification of an SF3B4 mutation.

Differential diagnosis may include mandibulo-facial dysostosis syndromes such as Treacher-Collins syndrome, and other acrofacialdysostoses (AFD) such as Catania-type AFD, Palagonia-type AFD, postaxial AFD (Genec-Wiedemann type), Rodriguez-type AFD, and mandibulo-facial dysostosis-microcephaly syndrome (see these terms). Patients with Goldenhar syndrome may also have overlapping features with NAFD.

Prenatal diagnosis can be performed by ultrasonography or by molecular test for SF3B4.

The clinical management for most cases mainly depends on a multidisciplinary approach consisting of maxillofacial radiology, paediatrics and neonatology, otolaryngology, anaesthesia, plastic surgery, oral and maxillofacial surgery

including cleft palate repair, mandibular lengthening with distraction osteogenesis or orthognathic surgery, among others, including bone grafts and ramus reconstruction [4]

In this clinical case, the patient opted for conventional treatment and refused any surgical intervention, the indication of extraction of the remaining maxillary teeth was necessary given the mobility and the advanced caries process and therefore a complete denture was performed to improve aesthetics and function of the patient.

Prosthetic rehabilitation was challenging due to multiple reasons; the micrognathia, skeletal Class III, deformation of palate and presence of cleftpalate, limited mouth opening and inadequate amount of supporting tissues.

The fabrication procedure of the denture was carefully made to achieve retention and stability; it significantly helps in re-establishing of the function and improves the patient's quality of life.

Conclusion:-

Conventional complete denture therapy with carefully recorded surface and balanced occlusal surface can provide favorable retention and stability even though the denture bearing tissues are unfavorable especially in craniofacial disorder patients.

References:-

1. Andrew M. Simpson, Sagar T. Mehta, Faizi Siddiqi, Duane Yamashiro, and BarbuGociman. (2018): Modified Lefort Distraction Osteogenesis for the Treatment of Nager Syndrome-Associated Midface Hypoplasia: Technique and Review. The Journal of Craniofacial Surgery., 29(6).
2. BozatIJoLlu R, MünevveroLlu A.P (2015): Dental Management of a Patient with NagerAcrofacialDysostosis. Hindawi Publishing Corporation Case Reports in Dentistry, ID 984732, pp 4
3. Hall B. (1989): Nageracrofacialdysostosis: autosomal dominant inheritance in mild to moderately affected mother and lethally affected phocomelic son. Am I Med Genet., 33pp.394-7.
4. Hari K, M Siva K, Vishalakshi S K, Sabitha H K (2015): Nageracrofacialdysostosis: a rare genetic disorder causing bilateral temperomandibular joint ankylosis in a 10-year-old girl. BMJ Case Rep. doi:10.1136/bcr-2015-212949
5. Kawira EL, Weaver DD, Bender HA. (1984): Acrofacialdysostosis with severe facial clefting and limb reduction. Am 7 Med Genet 17:641-7.
6. Krauss C, Hassell L, Gang D. (1985): Brief clinical report: anomalies in an infant with Nageracrofacialdysostosis. Am 7 Med Genet; 21:761-4.
7. Lowry RB. (1977): The Nager syndrome (acrofacialdysostosis): evidence for autosomal **dominant** inheritance. Birth Defects; 13:195-220.
8. Marie T McDonald, Jerome L Gorski (1993) Nageracrofacialdysostosis. J Med Genet 30: 779-782
9. Paul A. Trainor, and Brian T. Andrews (2013): Facial Dysostoses: Etiology, Pathogenesis and Management. Am J Med Genet C Semin Med Genet.163(4).
10. SaltajiH, Altalibi M, Major MP, et al. (2014): Le fort III distraction osteogenesis versus conventional le fort III osteotomy in correction of syndromic midfacial hypoplasia: a systematic review. J Oral MaxillofacSurg72:959–972
11. Shahin A F., NikzadS., Mehrnoush M. (2012): A Case Report: NagerAcrofacialDysostosis. Iranian Journal of Otorhinolaryngology 24(66).