ORAL REHABILITATION OF A PATIENT WITH ECTODERMIC DYSPLASIA SYNDROME

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ABSTRACT

Oral clinical manifestations of ectodermal dysplasia are in a wide variety, ranging from the presence of 2 or 4 to 6 teeth (oligodontia) usually with abnormal shape and size, with short, curved and thin roots to subtotal anodontia or total anodontia. There is no rule regarding missing teeth, nor the position or form under which they are found. Often the position of the teeth on the arcade is unfavorable, requiring a complex interdisciplinary treatment, involving the orthodontic repositioning first, followed by the prosthetic restoration through the infant prosthesis during childhood, and ultimately over-implant prosthetic restoration after the end of the growth process.

For oral rehabilitation it is essential to know the age of the patient, the number and condition of the teeth present, and the level of the growth process.A 4-year-old patient who visited our clinic was treated only prosthetically, because the clinical situation allowed us to address this treatment option. Infant prostheses were applied at this age, the final treatment plan having as objective the rehabilitation of all the functions affected by supraimplantation prostheses after the end of the growth processes.

Key words: ectodermal dysplasia, anodontia, oral rehabilitation, growth process.

INTRODUCTION

Ectodermal dysplasia is a rare, chromosomial X-linked condition. It appears in all races, with an incidence of 1 to 7 in every 100 000 births[1]. The syndrome presents itself as a triad of symptoms: hypo or anodontia, hypotrichosis[2] and hypohidrosis, and has associated problems that derive from the abnormal development of the ectodermic structures[3]. Patients with ectodermal dysplasia have prominent supraorbital ridges, frontal bossae and nose saddle. The nose itself is thin, pinched, with underdeveloped alae. An „elderly" look is observed, resembling the edentulous patient[4,5]. Other symptoms include hypolacrimation and hyposalivation, mucosa with functional problems, frequent respiratory infections, sight and hearing impediments, missing fingers and toes, cleft palates, immunitary deficiencies, sensitivity at exposure to light, underdevelopment of mammary glands[4,5,6].

Dental modifications include small, peg-shaped incisors. Both temporary and permanent teeth are involved. Anodontia can occur, but more frequently we encounter hypodontia with abnormally shaped remaining teeth, sometimes affected by hypoplasia[4,7].

MATERIAL AND METHODS

In this article we will try to describe a clinical case with subtotal anodontia that was diagnosed early during childhood, in the general context of the ectodermic dysplasia syndrome that was present.

Clinical case:

S.M., a 4 year old female, sought treatment in the Orthodontics clinic in Cluj-Napoca because of the functional and esthetic troubles of the dento-maxillary complex, both during rest and exerting function. The patient had been diagnosed with ectodermic dysplasia syndrome since the age of 3. The parents deny that any other member of the family suffers or suffered from the same disease.

Extraoral examinations shows symptoms consistent with ectodermal dysplasia syndrome: old-looking face, with everted lips, prominent chin, deep nasolabial and mental folds, thin hair follicles, pale skin (fig. 1).

Intraoral, we can see a dry oral mucosa, underdeveloped alveolar ridges in height and width. Teeth 5.5 and 6.1 are present in the upper arch, with 6.1 having a reduced size and an abnormal, peg-like shape. In the lower arch only 8.5 is present, having a normal shape and size (fig. 2) . A subtotal anodontia diagnosis was established. The clinical situation led to choosing of a prosthetic treatment plan: infantile dentures. Prosthetic treatment was carried out by following the usual stages of making the partial acrylic dentures: upper and lower preliminary impressions and casts, making of individual impression trays, functional impression, recording intermaxillary relations by using record bases and wax rims (fig. 3), fitting of the wax trial denture (fig. 4), leading to final placing of the denture in the patient's mouth (fig.5 and fig. 6).

Figure 1. Sm, 4 years old, extraoral view

Figure 2. Sm, 4 years old, intraoral situation

Figure 3. Recording of the intermaxillary relations

Figure 4. Wax trial dentures being adapted

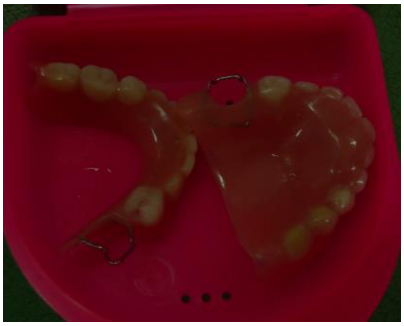

Figure 5. Final dentures

Figure 6. Final dentures in place

After placing the dentures, the patient came back for adjustments after 24 hours, a week, two weeks and after a month.

RESULTS AND DISCUSSIONS

Ectodermic dysplasia has early manifestations in the dento-maxillary area, starting with the temporary dentition period. Diagnosis and treatment is esential as fast as possible, any treatment having the purpose of reestablishing normal function and avoiding psycosocial complexes than can have their roots even at this age. In this certain case, relative easy prosthetics treatment was possible due to the normal position of the teeths, preprosthetic treatment not being necessary. The teeth that were present allowed retention of the dentures by means of Adams' clasps. The dentures reestablished normal function of the dento-maxillary complex, real benefits being brought to the patient, both from an esthetic and a functional point of view. We feel necessary to state that the dentures will be periodically replaced, at intervals of one and a half to two years, in order to allow development of the maxillary bones. Parents were made aware of this aspect and of the importance of regular visits. At the end of the growth period, the range of therapeutic means will increase, having the option of placing osseointegrated implants and an overdenture.

CONCLUSIONS

1. The rarity of this clinical situation, the major changes in the oral structures present in ectodermal dysplasia require interdisciplinary treatment since the temporary dentation period, in order to restore all dento-maxillary functions.

2. This creates the optimal conditions for the growth and development of the maxillary bones, which in the future fosters the application of long-lasting therapeutic solutions.

3. Although the clinical approach of the case is multidisciplinary, some patients can only be successfully treated through prosthetic treatment. Our case demonstrates, through the favorable clinical situation, this treatment option.

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