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CESE REPORT

Ectodermal dysplasia with anodontia – case report

Dysplazja ektodermalna z anodoncją – opis przypadku

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Abstract

Ectodermal dysplasia is caused by genetic anomalies, which can be inherited through parents or gene mutations. Both modes of inheritance (recessive and dominant) have been described. The X-linked pattern of inheritance is also possible. Ectodermal dysplasia is characterized by abnormalities in the tissues of ectodermal origin. The anomalies can be particularly evident in skin, nails, hair and teeth. ***Aim of the study****. To present a case report of a child suffering from ectodermal dysplasia with total anodontia.* ***Materials and method****. The study presents prosthetic treatment of a 3-year-old male patient with anhydrotic ectodermal dysplasia and complete anodontia. The treatment of choice was preparation of complete maxillary and mandibular dentures. This method provided proper development of mastication, speech and acceptable appearance. Complete dentures were prepared. Instructions on the use of prostheses and oral hygiene maintenance were given to the parents.* ***Results****. After a few weeks, the boy was completely adapted to using the dentures.* ***Conclusion****. Clinical knowledge of craniofacial development and growth is necessary for the treatment of patients with ectodermal dysplasia. The multidisciplinary team approach is required.*

Streszczenie

Dysplazja ektodermalna jest zaburzeniem genetycznym mogącym mieć charakter dziedziczny lub powodowanym mutacją genetyczną. Może być dziedziczona w sposób recesywny i dominujący. Może być również dziedziczona w sposób związany z chromosomem X. Dysplazja ektodermalna charakteryzuje się zaburzeniami tkanek pochodzenia ektodermalnego. Zaburzenia dotyczą skóry, paznokci, włosów i zębów. ***Cel pracy.*** *Celem pracy był opis przypadku dziecka chorego na dysplazję ektodermalną z występującą anodoncją.* ***Materiał i metody.*** *Praca przedstawia przebieg leczenia protetycznego dziecka chorego na anhydrotyczną dysplazję ektodermalną, z całkowitą anodoncją. Plan leczenia pacjenta obejmował wykonanie całkowitej protezy górnej i dolnej. Umożliwiło to prawidłowy rozwój żucia, mowy oraz akceptację wyglądu. Wykonano protezę całkowitą górną i dolną.. Rodzice dostali wytyczne dotyczące użytkowania uzupełnień oraz prawidłowego przeprowadzania zabiegów higienicznych w obrębie jamy ustnej.* ***Wyniki.*** *Po kilku tygodniach użytkowania chłopiec całkowicie zaadoptował się do uzupełnień protetycznych.* ***Podsumowanie.*** *Leczenie pacjentów chorych na dysplazję ektodermalną wymaga wiedzy klinicznej dotyczącej rozwoju i wzrostu twarzoczaszki. Konieczne jest leczenie multidyscyplinarne.*

KEYWORDS:
ectodermal dysplasia, anodontia, children, prosthetic treatment

HASŁA INDEKSOWE:
dysplazja ektodermalna, anodoncja, dzieci, leczenie protetyczne

Introduction

Ectodermal dysplasia is a congenital syndrome. It is characterized by abnormalities in the ectoderm developing tissues. The anomalies can be particularly evident in skin, nails, hair and teeth.[1,2]

Ectodermal dysplasia is caused by genetic anomalies, which can be inherited through parents or gene mutations.[1] Ectodyspalsin (EDAI), EDA receptor (EDAR) and EDAR associated death domains (EDARADD) as genes responsible for changes have been described. Both modes of inheritance (recessive and dominant) are possible. The X-linked pattern of inheritance is also possible.[3]

Ectodermal dysplasia can be classified into hypohidrotic and hidrotic. Hypohidrotic type is the more severe form. It is associated with sensitivity to heat, frequent high fevers and dento-facial anomalies.[1,2] In hidrotic type, sweat glands are normal.[4]

Most people with ectodermal dysplasia have apparent facial asymmetry which includes frontal bossing, depressed nasal bridge, vertical facial height and depth, small palatal and cranial base widths, small malar processes and high-set orbits. Hypodontia, or even anodontia, is a characteristic occurrence, which can lead to deepening the cranio-facial asymmetry. The atrophy of alveolar process in the edentulous region may cause reduction in the lower facial heigh.[1-3,5,6]

The clinical knowledge of craniofacial development and growth is necessary for the treatment of patients with ectodermal dysplasia. Patients in young age usually present a multitude of anomalies. That is why a multidisciplinary team approach is required.[1]

The whole plan of dental treatment should be created together with a child and parents and have their full acceptance. Oral rehabilitation of patients with ectodermal dysplasia usually consists in preparation of removable prostheses. It is advisable to have the initial prosthesis ready before a child starts school for easier adaptation and getting used to the denture.[1,7]

Early initiation of prosthetic treatment increases the tonsity of masticatory muscles, retards the resorption of alveolar bone caused by the absence of teeth, and compensates the reduction in vertical dimension.[1,8] It has a significant impact on the developmental process of mastication and speech. It results in self-acceptance and acceptance by peers.[1]

Aim of the study

The study presents a case report of a 3-year-old male patient with hypohidrotic ectodermal dysplasia and complete anodontia.

Material and method

A 3-year-old boy was referred to a private dental clinic (Fig. 1). The reason for the patient's visit was lack of teeth and problems with speech and mastication. The patient was diagnosed with anhidrotic ectodermal dysplasia in Children's Hospital in Lublin, where he was treated. His brother and sister were healthy. Parents also did not have symptoms and there was no other family history of ectodermal dysplasia.

Extraoral examination showed features typical of ectodermal dysplasia such as extremely wispy, sparse hair, saddle nose, dry and light-coloured skin and thin wrinkles around the mouth. Intraoral examination did not reveal any erupted teeth. Alveolar ridges were thin. There was reduced vertical high of alveolar bone.

Complete anodontia was confirmed by panoramic radiography (Fig. 2). On radiographs there were no permanent or deciduous tooth germs.

The best treatment of choice was with complete maxillary and mandibular dentures. This method provided proper development of mastication, speech and acceptable appearance.

Plan of treatment was explained to the parents. Routine procedures were followed for the construction of dentures. Preliminary impressions were made and then the custom trays for functional impressions were used. Functional impressions were made with c-silicone impression materials of two different densities (Fig. 3). On the basis of functional impressions plaster models were made (Fig. 4).

The maxillary and mandibular rims were adjusted. The maxillo-mandibular relations were established and were further transferred to the

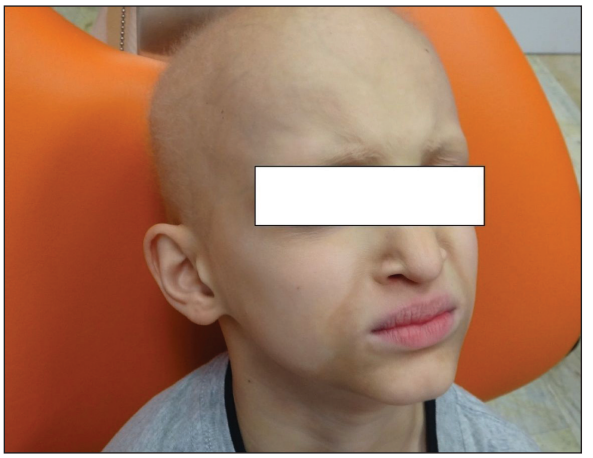

Fig. 1. *Patient with Hypohidrotic Ectodermal Dysplasia – present moment. Now the patient is 5 years old.*

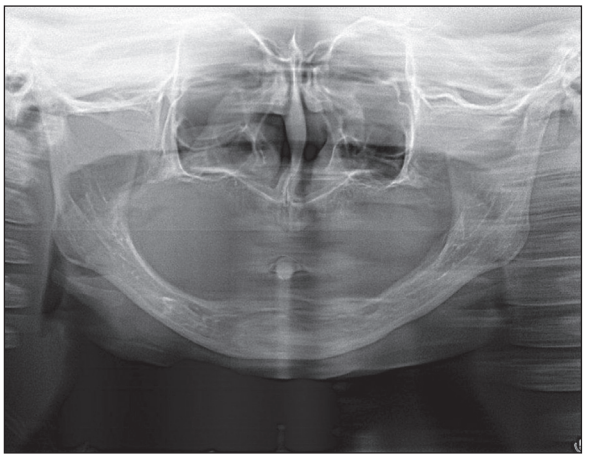

Fig. 2. *Panoramic radiograph – complete absence of teeth.*

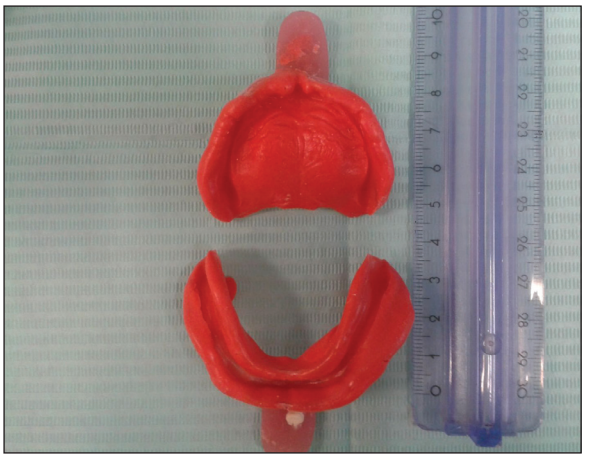

Fig. 3. *Functional impressions.*

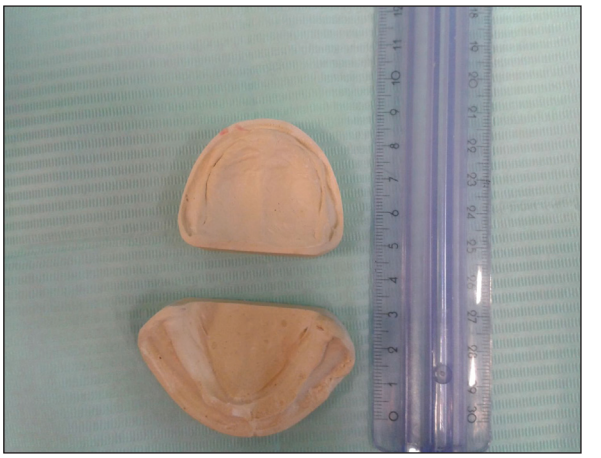

Fig. 4. *Plaster models.*

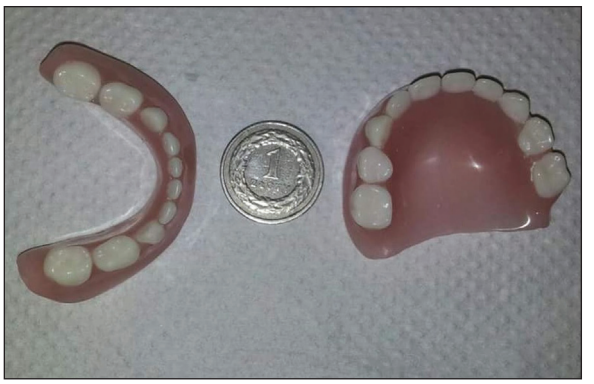

Fig. 5. *Complete dentures.*

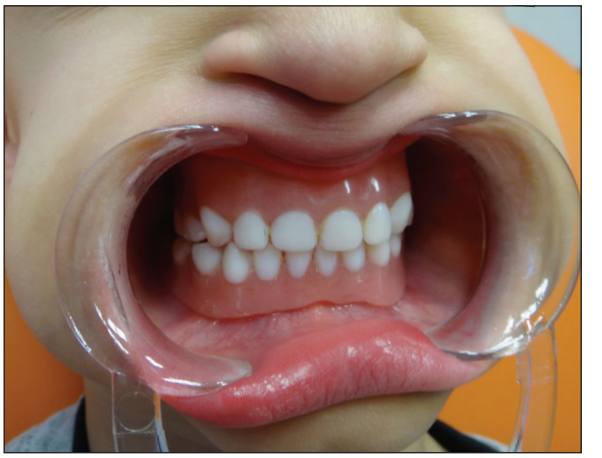

Fig. 6. *Patient with his dentures.*

articulator. Primary tooth forms were selected. Try in was done and the child and the parents were satisfied. During the next visit the patient got complete dentures (Fig. 5, 6). Oral hygiene instructions were given to the parents.

After a few weeks, the boy completely adapted to using the dentures. Good retention was observed.

To ensure proper growth and development further recall appointments were scheduled every three months. During those visits, the interferences were eliminated, rebasing was made. Until today, new dentures were made twice: after one year and after nine months. The dentures provided proper conditions for the maintenance of a normal diet and acceptance among peers. It was possible to improve development of speech and communication skills.

Discussion

Treatment of children with ectodermal dysplasia is very complicated. The disease may have various manifestations. The additional difficulty is dealing with a child patient and its problems. Prosthetic treatment is required for the proper development and functioning of the stomatognathic system. It also affects the psychological development of a child and the acceptance of their peers, thereby improving the quality of life of the patient.[9,10]

Some clinicians suggest that prosthetic treatment should be initiated at the age of 5 years. The patient described in this paper got his first denture at the age of three years and after a few weeks complete adaptation was noted.

Early rehabilitation of anodontia ensures proper tonicity of masticatory muscles and compensates the decrease of vertical dimension. Early treatment with complete dentures leads to significant improvement in mastication, development of speech and satisfactory diet.[1,11]

The patient's parents reported significant improvement in speech and social adaptation.

The cooperation of parents is essential to the success of prosthetic treatment of a child.[1] This patient had full support, especially from his mother. This had a significant impact on adaptation to dentures and acceptance of the ectodermal dysplasia symptoms.

Control visits are necessary and important; they are aimed at modification or replacement of the denture due to changes related to the development and growth.[1,7,11]

Summary

The symptoms of Ectodermal dysplasia cause significant problems in the development of stomatognathic system, speech, mastication and socialization. Patients can have problems with acceptation by peers. Early management of the disease influences proper development and significantly improves the quality of life in individuals affected by the condition.

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