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Chair and Department of Paedodontics Dentistry, Medical University of Lublin

MARIA MIELNIK-BŁASZCZAK, MAREK TOMANKIEWICZ

A rare case of tooth-nail syndrome

Ectodermal dysplasia is a general term describing distempers of ectodermal tissue organs, such as skin and appendages, teeth, nails, etc. At present we can differentiate 150 types of it classified in 11 subgroups on the basis of their clinical manifestation or etiology. These syndromes are inherited, with the X-chromosome as an autosomal dominant or recessive genetic trait (5).

CASE DESCRIPTION

A female patient J. K., aged 12, was first examined at the Department of Developmental Age Dentistry at the Medical University of Lublin in April 2002. The extraoral examination showed the proper structure and symmetry of the face (facial profile also proved the symmetry of facial features), the skin was of appropriate colour, there were no signs of rash. Trigeminal nerve ending was not painful; cervical and submadnibular lymph nodes were regular. Hand skin was dry; fingernails were of irregular shape. Body weight was 40 kg, height – 151 cm.

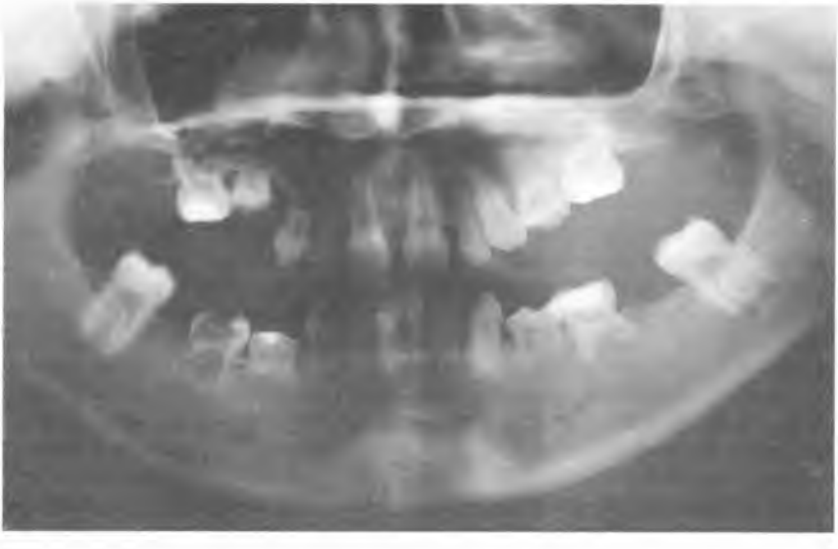
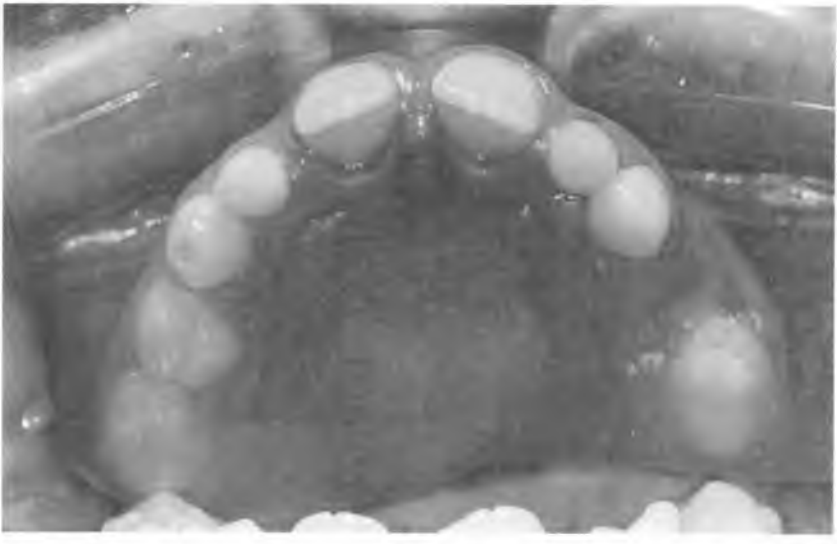
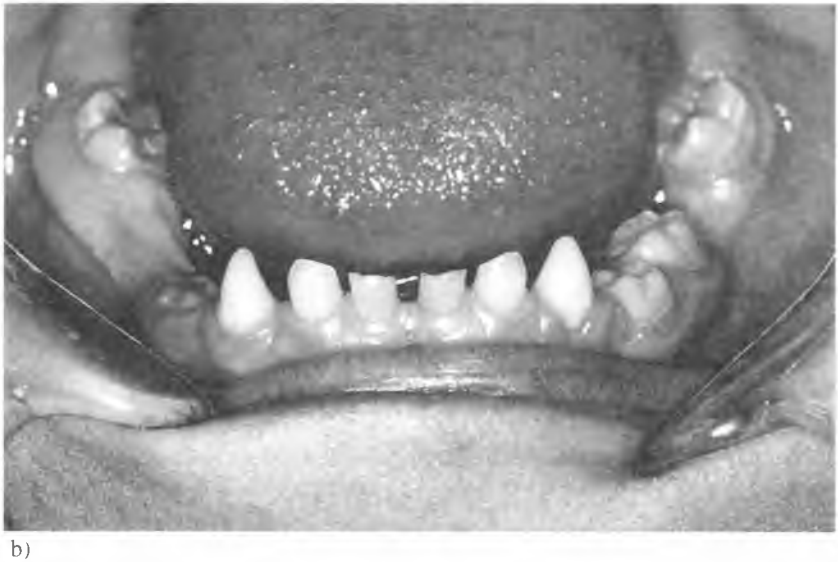


Fig. 1. The patient's panthomogram



a)



b)

Fig. 2. The patient's upper and lower tooth arch

Fig. 3.The finger nails

From the medical history of the patient we learnt that the pregnancy period was not disturbed in any way, the mother did not take any medicines. The labour was spontaneous, without any complications. The mother admitted that the infant did not have any fingernails. The child developed correctly. She suffered from mumps, chickenpox and rubella. All vaccinations were administered according to the schedule. Primary teething began normally; all primary teeth appeared.

During the intraoral examination it was discovered that the colour of the mucous membrane of cheeks and lips was normal, the marginal paradontium did not show any pathologies; the openings of the parotic gland were slightly bigger than normal. The examination showed the presence of numerous primary teeth; only teeth no 11, 21, 36 and 46 were secondary. The crowns of teeth 53, 63, 73 and 83 were irregular (Figs. 2a i 2b). Teeth number 54, 55, 64, 74, 84 underwent reinclusion (85 were removed because of chronic inflammation peripapical tissues). An additional radiological examination (panthomogram) showed numerous lacks of secondary teeth buds (Fig. 1). Endocrinologists did not observe any calcium or phosphorus disturbances. The results of morphological and biochemical blood tests or thyroid hormone level test were normal. On the basis of an X-ray of a wrist, the bone age was established to be 12 years (the same as the patient actually was). Dermatological examination (especially of the walls of nails) showed the presence of numerous bacteria. Capillaroscopy showed the presence of numerous capillary loops at the walls of fingernails, the background was pink and red (Fig. 3). Ophthalmologists did not observe any malfunctioning of the fundus and front chamber of the eye. On the basis of the results of the above-mentioned examinations a diagnosis was formulated: the patient suffered from tooth-nail syndrome.

DISCUSSION

Tooth-nail syndrome was first described by Witkop. It usually affects teeth and nails (nails of infants are underdeveloped, it applies in particular to toenails). There may be no secondary teeth buds (it pertains to incisors, canine and premolar teeth of the mandibula and

canine teeth of the maxilla). Deciduous teeth might be of a conical shape. That is a feature that differs Witkop's Syndrome from hydrotic ectodermal dysplasia, where the number of teeth is not reduced. Contrary to anhydrotic ectodermal dyspalsia, Witkop's Syndrome is inherited as an autosomal dominant genetic trait; skin appendages are affected by the illness to a minimum extent (hair and sweat glands) (3). Eyebrows and eyelashes may be regular, head hair may be thinner or regular as well, ovarian cysts may appear (1,6).

Experiments on mice demonstrated that MSX1 gene is responsible for developing Witkop's Syndrome. Its deficiency caused the appearance of hypodontia in mice as well as disturbances in mesenchyme forming nail plates (4).

The dental treatment of such patients depends on the scale of dental deficiencies. Artificial denture seems to be necessary from the very early age in order to ensure proper chewing. Systematic changes of prosthetic appliances should be carried out until the end of puberty period. If possible, implants can be used to provide a final prosthetic appliance (2).

CONCLUSIONS

Patients with any form of ectodermal dysplasia should remain under permanent dental care since an early age. It makes it possible to ensure proper chewing and reduce the discomfort of a patient.

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SUMMARY

The present work is a case report of a 11-year-old patient directed for consultations to Department of Paedodontics, Medical University of Lublin. The clinical examination ascertained numerous lacks of permanent teeth, there were only central incisors in maxilla and first molars in mandibula. With the exception of teeth 54, 55, 64, 74, 84, which were reincluded, the patient had remaining deciduous teeth. The radiographic examination confirmed large oligodontia within permanent teeth. Hair, sweat glands, anterior chamber and bottom of the eye were correct. Little disturbances within nail plates of palm fingers were found. A gentle form of ectodermal dysplasia was recognized as a tooth-nail syndrome (Witkop's syndrome).

Rzadki przypadek zespołu zębowo-paznokciowego

Praca jest opisem przypadku 11-letniej pacjentki, skierowanej na konsultacje do Katedry i Zakładu Stomatologii Wieku Rozwojowego AM w Lublinie. Badaniem klinicznym jamy ustnej stwierdzono liczne braki zębów stałych, obecne były jedynie siekacze centralne w szczęce i pierwsze stałe trzonowce w żuchwie. Z wyjątkiem zębów 54, 55, 64, 74, 84, które uległy reinkluzji, pacjentka posiadała wszystkie pozostałe zęby mleczne. Wykonane badanie pantomograficzne potwierdziło podejrzenie dużej oligodoncji w obrębie zębów stałych. Włosy, gruczoły potowe, przednia komora i dno oka były prawidłowe. Wykazano niewielkie zaburzenia w obrębie płytek paznokciowych palców dłoni. Na podstawie badania klinicznego oraz zebranego wywiadu i badania ogólnolekarskiego rozpoznano łagodną postać dysplazji ektodermalnej w postaci zespołu zębowo-paznokciowego (zespół Witkopa).