Case Report

Hidrotic ectodermal dysplasia in a 40-year-old female patient

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BACKGROUND: Hidrotic ectodermal dysplasia presents with anomalies such as enamel hypoplasia, hypodontia and facial dysmorphy. In the developed world diagnosis is done during childhood, but in the developing countries patients fail to report for various reasons. Continued documentation of such conditions therefore remains important in oral health. **AIM:** To report a case of hidrotic ectodermal dysplasis that was successfully treated with removable dentures. **FINDINGS:** This patient was assessed clinically and by the use of X-ray. The patient presented with mild dysmorphy, severe enamel hypoplasia and hypodontia. The radiograph revealed retained teeth at different stages of development. **DISCUSSION:** The patient was disturbed by her appearance and pain that prompted her to seek medical attention. Such cases may exist in society but probably resort to traditional healers thus remains undocumented. **CONCLUSION:** Oral health education is important to bring awareness regarding different conditions of the orofacial region and the importance of seeking medical attention.

Key words: Esthetic, function, hidrotic ectodermal dysplasia

INTRODUCTION

Enamel hypoplasia, hypodontia and facial dysmorphy are among conditions grouped together under a rare congenital syndrome known as ectodermal dyslasia (ED), which affect tissues of ectodermal origin such as teeth, hair, nails, skin and sweat glands.[1,2] There are two types of ED classified on basis of presence or absence of sweating. In anhidrotic ectodermal dysplasia (EDA) type, sweating is reduced or absent while in hidrotic ectodermal dysplasia (EDH) type, sweating is not affected.[3] ED is also associated with hypoplasitic or severely malformed teeth, retained deciduous and missing permanent teeth.[4,5] In the EDA type the patient may also present with shiny skin and alopecia of varying degrees.[2] In the developed world ED are diagnosed based on laboratory identification of genes associated with ED[6,7] and mode of inheritance of the mutant genes have been suggested to be either recessive X chromosome or autosomal dominant and recessive.[2,8] This may be difficult in the developing societies where such facilities and oral health services are insufficient. The current observation report a case of EDH based on clinical presentation, family history and X-ray investigation.

Past medical and family history

Patient had no history of major or chronic illnesses. She reported normal sweating. Patient reported to have a young sister who had a similar condition, although it was not documented. She also reported her mother's concern about the patient's teeth when she was young but had associated the deformity of the deciduous teeth with drugs she was given during her pregnancy.

Past dental history

She gave a history of deciduous teeth deformity. Some of the teeth were normally shed but there was no eruption of successors and some were retained. She also reported the condition got worse with time, some teeth getting mobile and painful, while some underwent severe attrition leaving root stumps.

CASE REPORT

The present work is a report on a 40-year-old lady who came with a chief complaint of deformed and missing teeth since childhood. Of recent she was concerned with pain, badly malformed teeth, which seem to get worse with time and the facial appearance. The patient thus requested for extraction of all the teeth because she was not happy with them and they were not very useful in chewing.

On general examination there was a slight facial dysmorphy with midline shift to the right [Figure 1]. No other abnormalities were noted on the head and

neck. Ears were normal and she looked healthy with normal hair and skin. Examination of the fingers and toes revealed normal nails.

Intra-oral examination revealed both jaws with few deformed permanent and deciduous teeth and some root remnants [Figure 2]. Most teeth were missing and the few present were severely hypoplastic and mobile and some root stumps were also observed. Major parts of the alveolar ridges were normal except resorption of the upper and lower alveolar ridges at areas corresponding to 13 and 46, respectively [Figure 3].

Management

Radiological investigation using an orthopantomography was done following general and intra oral examination. The radiograph revealed missing and submerged teeth and ruled out bone deformity [Figure 4]. All the teeth and roots were planned for extraction and non-erupted teeth left in place. Full blood picture and differential cell counts was requested and results were essentially normal. This was to rule out T cell immunodeficiency, which is commonly associated with anhidrotic ectodermal dysplasia.[9]

Canceling was done to explain to the patient about her condition, how it is acquired, possible treatment modalities and post treatment care.

Extraction of teeth was done at four sittings, each time a quadrant was done and given a few days healing time. A broad-spectrum antibiotic was prescribed to control secondary infection. The patient was requested to stay for a week following the last extraction session to give time for complete wound healing.

Thereafter the negative impressions were taken for preparation of positive models and fabrication of acrylic removable full dentures.

The patient was satisfied with her appearance and pain relief and was very motivated and hoped for functional improvement with the dentures in place.

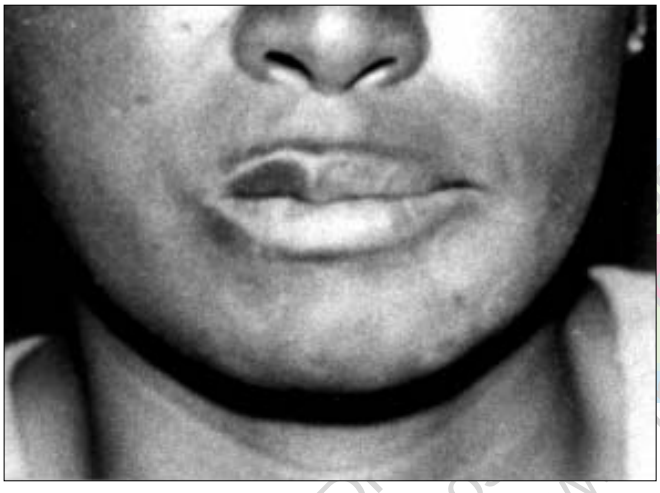

Figure 1: A photograph of the lower part of the face showing the median plane shift to the right

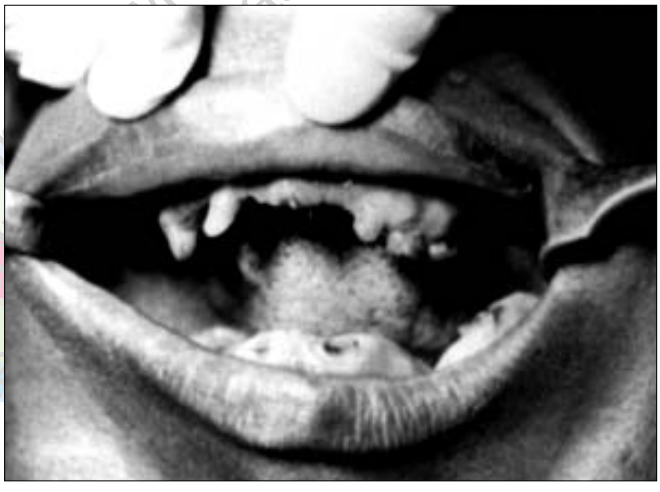

Figure 3: An intra-oral photograph to display the hypoplastic teeth and root remnants

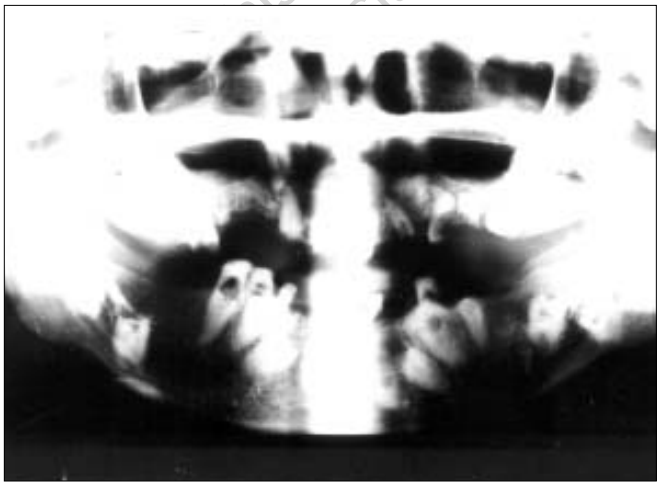

Figure 2: A photograph of an orthopantomography radiological film showing retained teeth with full-formed crowns. Roots could not be observed

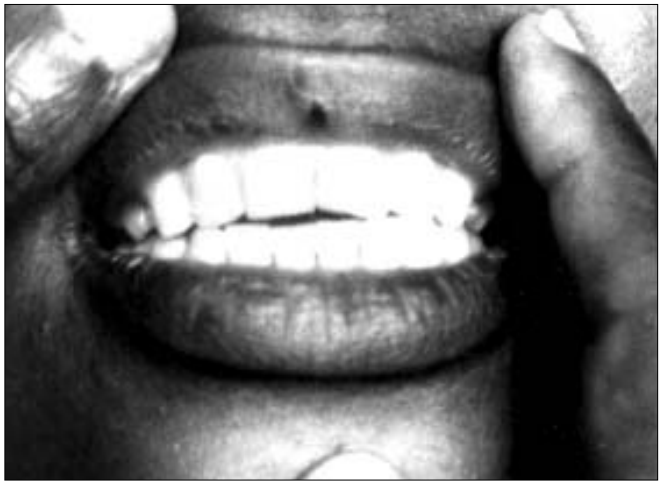

Figure 4: An intra-oral photograph showing the fabricated acrylic removable dentures won by the patient

Patient was discharged and asked to report to the clinic after 4 weeks for first evaluation mainly on expected changes after complete bone healing. The patient reported back after the first 4 weeks of using her new set of teeth and was happy with the outcome.

DISCUSSION

This report regards an adult female patient with EDH who visited the restorative clinic of the School of Dentistry with features suggestive of ED. The patient presented with pain, severely hypoplastic and missing teeth, accompanied with facial asymmetry. It is the first case of its kind to be documented in Tanzania. This can be explained by possible lack of correct information and knowledge, resulting in lack of awareness. Such patients therefore may seek treatment elsewhere such as from traditional healers and may live with such deformities for life. Although in this case the disabilities were limited to teeth and face ED may cause different forms of disabilities such as different degrees of alopecia and anodontia and severe disabilities such as immunodeficiency and hypohidrosis or unhidrosis, which may require continuous palliative management.[7] Thus it may pose serious health problems especially in some societies where marriages within families (such as between cousins) are practiced.[5,6] It is therefore important to give correct information regarding causes, disabilities associated with ED and management options to the professionals and society. In the current case the use of removable acrylic dentures following total extraction offered of teeth solved the patient's complaints by removing pain and improving patient's appearance.

The diagnosis of this case of EDH, reported late in life, was done based on clinical presentation and was managed successfully by cancelling the patient and providing her with acrylic removable dentures following total extraction. It is recommended that continued documentation and oral health campaigns to be conducted to raise awareness among the professionals and general population on common and rare conditions of the orofacial region for early reporting and management.

ACKNOWLEDGEMENT

The patient gave consent for intra and extra oral pictures and their publication, but with a request that her face not to be displayed in a way that can easily be identified when published. We thank the patient for her cooperation and consenting for her pictures to be used in this publication.

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