PICTORIAL

HYPOHIDROTIC ECTODERMAL DYSPLASIA

Manas Bajpai, Nilesh Pardhe
Department of Oral and Maxillofacial Pathology, NIMS Dental College, Jaipur, Rajasthan-India

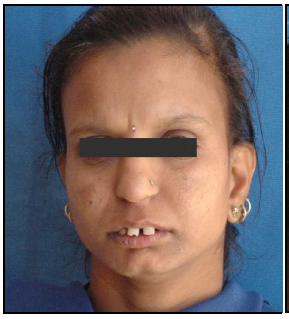

Figure-1: Front view of patient shows sparse hair, absence of eyelashes and eyebrows with averted lips

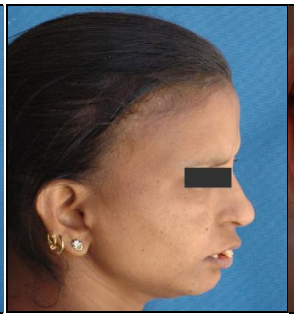

Figure-2: Side profile of patient

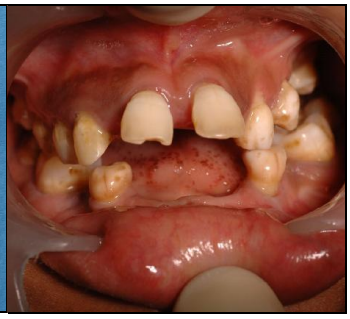

Figure-3: Intra oral picture of patient shows conical central incisors and missing mandibular central incisors

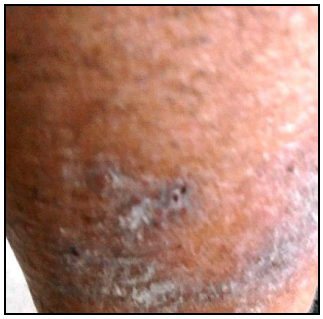

Figure-4: Eczematous lesions on lower extremity of the same patient

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A 23 year old lady reported with the chief complains of missing teeth. She had been diagnosed much earlier by the physicians for Ectodermal dysplasia. General examination revealed sparse hair, absence of eyelashes and eyebrows with frontal bossing and averted lips. The skin was dry and wrinkled. She had numerous eczematous lesions in lower extremities. There was a history of reduced sweating and heat intolerance. Intra oral examination revealed conical shaped maxillary incisors and missing mandibular central incisors. The teeth were chalky white in colour showed typical features of enamel hypoplasia. A generalized spacing between teeth was found. Panoramic radiograph showed compete absence of mandibular incisors with improper radiodensity between enamel and dentin.

Hypohidrotic ectodermal dysplasia is a rare genetic disorder characterized by the faulty development of the ectodermal structure, resulting in most notably anhydrosis/hypohydrosis, hypotrichosis and hypodontia. The condition is usually an X-linked recessive disorder affecting predominantly males.[1]

REFERENCES

1. De Aquino SN, Paranaíba LMR, Swerts MSO, Martelli DRB, de Barros LM, Júnior HM. Orofacial Features of Hypohidrotic Ectodermal Dysplasia. Head and Neck Pathology. 2012;6(4):460-466. doi:10.1007/s12105-012-0349-4.

Address for Correspondence:

Dr. Manas Bajpai, Department of Oral and Maxillofacial Pathology, NIMS Dental College, Shobha Nagar, Jaipur-Delhi Highway (NH-11C), Jaipur-303121, Rajasthan-India
Tel: +91 9799415000
Email: dr.manasbajpai@gmail.com