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.

REFERENCES

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