The Ectodermal Dysplasia!

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Introduction

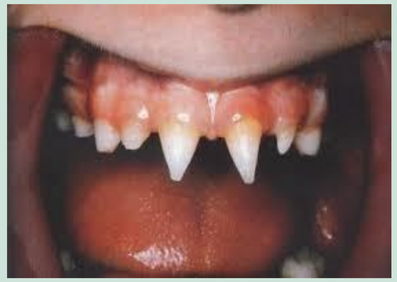
The Ectodermal Dysplasia (ED) is a genetic disorder (X-linked recessive) affecting the development or function of the embryonic ectoderm from which the teeth, hair, nails and sweat glands develop. Ectodermal Dysplasia (ED) is not a single disorder, but a group of closely related conditions of which more than 150 different syndromes have been identified[1].

Genetic pathogenesis

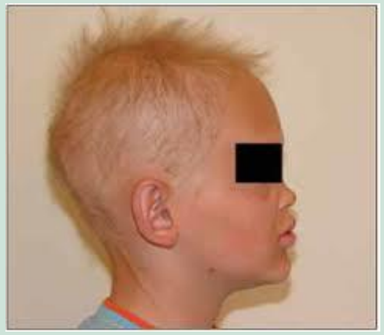
The EDA, EDAR and EDARADD genes provide instructions for making proteins (ectodysplasin A). that work together during embryonic development, Ectodysplasin A forms a part of a signaling pathway that is critical for the interaction between two cell layers, the ectoderm and the mesoderm. Mutations in those genes results in defective ectodysplasin A formation thereby preventing normal interactions between the ectoderm and the mesoderm[2] .

Clinical features

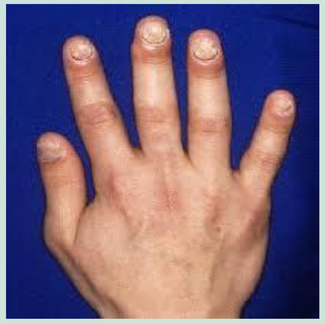
Ectodermal dysplasia can cause trichodysplasia, dental dysplasia, onychodysplasia, and dyshidrosis. This can be shown in the following figures[3].



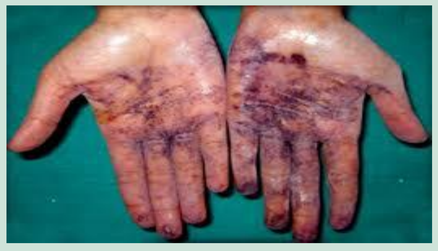
Dental dysplasia



Trichodysplasia



Onychodysplasia



Dyshidrosis

Conclusion

Ectodermal dysplasia is a rare genetic disorder with the involvement of various tissues in the body. A careful and a thorough examination of a patient will lead to an accurate diagnosis. It should be noted that an absence of a positive family history for ectodermal dysplasia should not be a factor in causing any diagnostic dilemmas with respect to ectodermal dysplasia, a condition that shows multiple modes of inheritance.

References

1. Deshmukh S, Prashanth S. Ectodermal Dysplasia: A Genetic Review. Int J Clin Pediatr Dent 2012; 5(3):197-202.
2. Wright JT., Grange DK., Ritcher MK. Hypohidrotic ectodermal dysplasia. Gene Reviews. NCBI BookShelf. Updated. 2009
3. http://www.skincareguide.ca/glossary/h/hypohidrotic_ectodermal_dysplasia.html