Salivary Gland Hypoplasia and Aplasia in Persons with Oligodontia and Ectodermal Dysplasias – Two Case Reports

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Introduction

Ectodermal dysplasias (EDs) comprise a large group of clinically and genetically heterogeneous conditions characterized by developmental defects in tissues derived from ectoderm. According to the literature, the most commonly affected tissues are hair, teeth, nails and sweat glands. Also the salivary glands have ectodermal origin. Salivary gland hypoplasia and aplasia in a few persons with hypohidrotic ED have been reported (Singh and Warnakulasuriya, 2004; Nordgarden et al, 1998), but have been referred to as rare manifestations X-linked hypohidrotic ectodermal dysplasia (XHED).

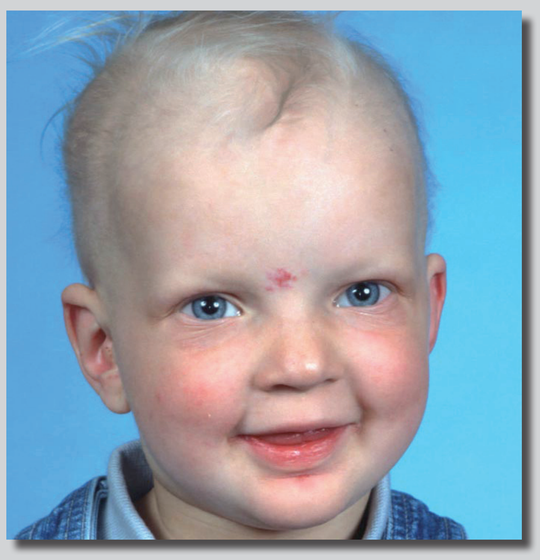
However, a few clinical studies have shown that salivary secretory rates is reduced in many persons with different forms of EDs, suggesting that salivary gland developmental disturbances are more common than described previously (Nordgarden et al, 2001; Bergendal et al, 2006). Here, two cases will be presented.

Case 1

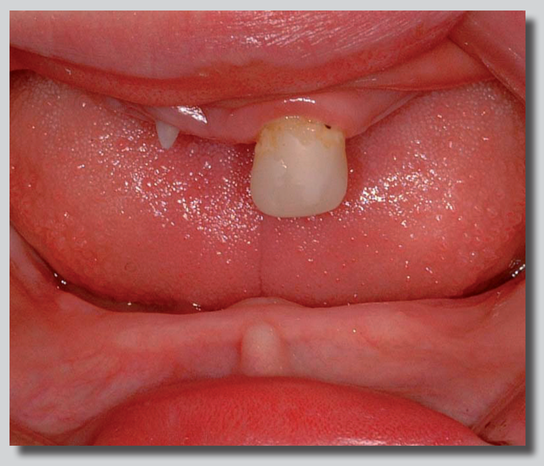
Ten year old male with XHED affecting hair, teeth and sweat glands. He demonstrated clinical signs of dry mouth.

As computer tomography (CT) was needed for dental treatment planning the salivary glands were examined at the same time.

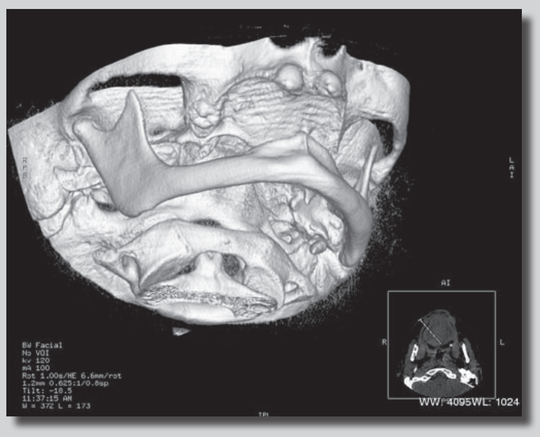
The CT scan revealed that his parotid glands were hypoplastic, and his submandibular glands could not be visualised at all.



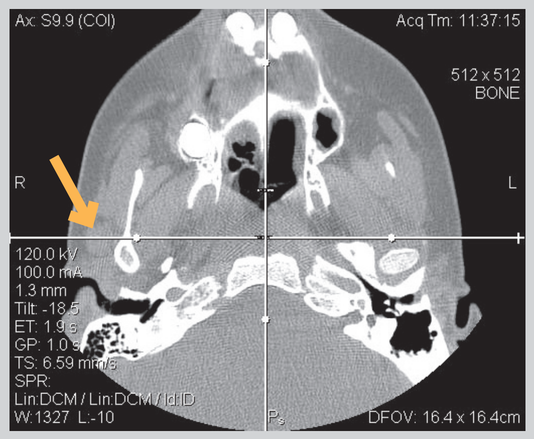
Case 1: At three years of age, he demonstrated typical manifestations of X-linked hypohidrotic ectodermal dysplasia.



Case 1: Oral status at the age of 8 years.



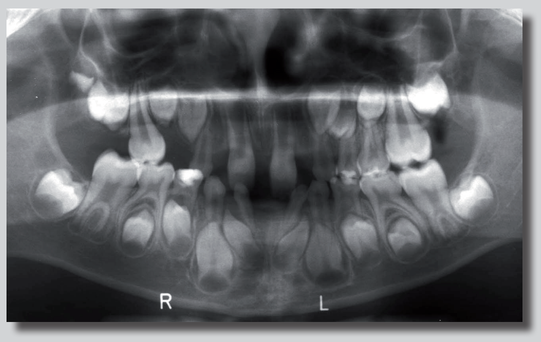
Case 1: Reformatted 3D image showing that only three permanent teeth are present and the resulting small and hypoplastic jawbones.



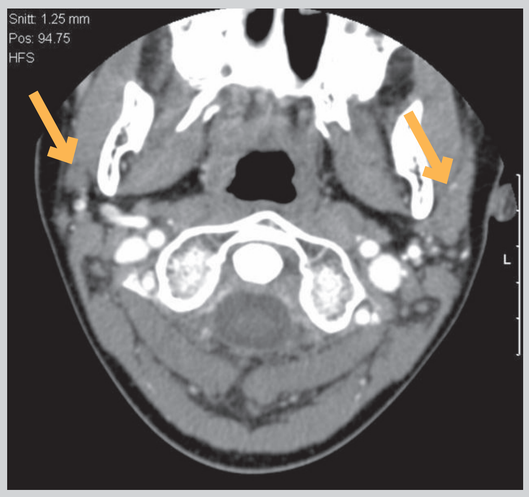
Case 1: CT-scan demonstrating hypoplastic parotid glands.

Case 2

Nine year old male with peg shaped incisors and canines, agenesis of five permanent teeth, taurodontia, delayed tooth development and high caries activity. His sweat capacity is not reduced, and other clinical symptoms and findings is not suggestive of an XHED. Hair and nails are normal. His oral mucosa seemed dry and he also reported a subjective feeling of dry mouth. CT demonstrated that both parotid and both submandibular glands were hypoplastic.



Case 2: Orthopantomogram at the age of 8 years demonstrating agenesis of one premolar and both lateral incisors in the upper jaw and two incisors in the lower jaw.



Case 2: CT-scan demonstrating hypoplastic parotid glands.

Conclusion

It is necessary to evaluate salivary gland function in person with several congenitally missing teeth with or without symptoms from other ectodermal tissues.

Persons with oligodontia may also have developmental disturbances of salivary glands, a condition we have chosen to term oral ectodermal dysplasia.

References

Bergendal B, Norderyd J, Bagesund M, Holst A. (2006). *Int J Paediatr Dent*, 16:320-6.
Nordgarden H, Jensen JL, Storhaug K. (2001). *Oral Dis*, 7:226-32
Nordgarden H, Johannessen S, Storhaug K, Jensen JL. (1998). *Oral Dis*, 4:152-4.
Singh P, Warnakulasuriya S. (2004). *J Oral Pathol Med*, 33:634-6.