Dental and oral lesions associated with focal dermal hypoplasia in two patients

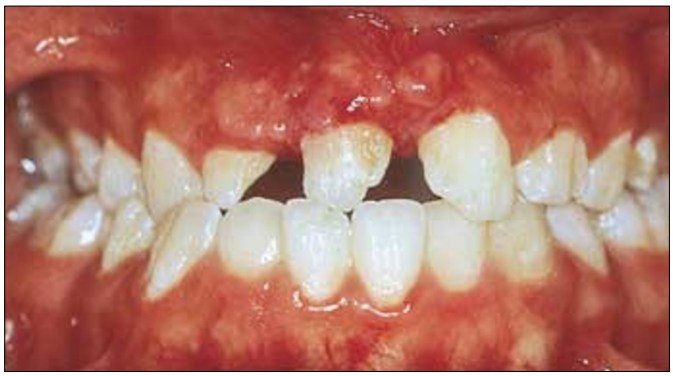
Dental and oral lesions in two patients with focal dermal hypoplasia (Goltz syndrome) by A. M. Baxter, M. J. Shaw, and K. Warren Br Dent J 2000; 189: 550-553

Abstract

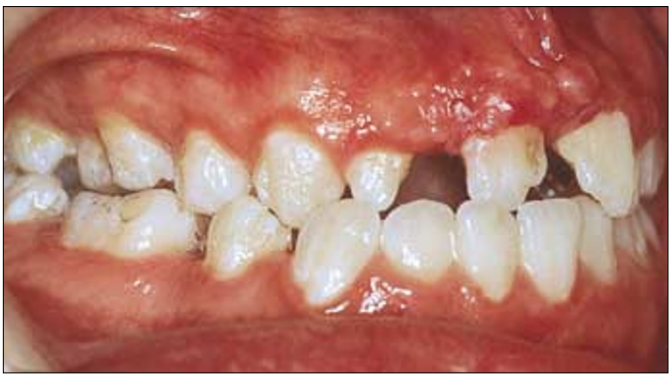
This report is concerned with the manifestations associated with focal dermal hypoplasia (Goltz syndrome) in two patients. Goltz syndrome is a rare polydysplastic syndrome. Systemic findings were similar to those previously reported in the literature and are illustrated. More detailed descriptions of the dental abnormalities are presented. The possibility of root resorption being, a previously unreported, part of the syndrome is hypothesised.

In Brief

- Focal dermal hypoplasia is a rare syndrome, which may have a profound effect on the oral and dental tissues.
- Careful clinical and radiographic examination will be helpful in defining the features of this syndrome.
- Although this is an uncommon syndrome the dental practitioner should be aware that external root resorption may be associated with the condition.
- Early diagnosis of external root resorption may help to avoid unnecessary tooth loss.
- The aetiology and classification of external root resorption is discussed.



Enamel hypoplasia and peg-shaped maxillary anterior teeth



Arborescent hyperplastic lesions affecting the labial gingiva related to the maxillary right central incisor tooth

Comment

Focal dermal hypoplasia is a rare developmental disorder affecting skin, eyes, teeth and the musculoskeletal system. This is now the preferred term for the syndrome, which has previously been known as Golz-Gorlin syndrome. The latter term could be confused with Gorlin-Golz syndrome, which is characterised by naevoid basal cell carcinomas, odontogenic keratocysts, skeletal abnormalities and intracranial calcifications.

Baxter, Shaw and Warren present two cases with focal dermal hypoplasia. Both cases exhibited several of the classical features of this rare syndrome — disorder of skin pigmentation, hand and finger defects, micropthalmia and skeletal abnormalities of the spine. In addition, both cases had dental features which have been previously associated with the condition — enamel hypoplasia and hypodontia. On radiographic examination, external root resorption was evident in a central incisor in one case and in a first permanent molar in the other.

The dental management of Case 1 is described in detail. This included cosmetic improvement of the anterior teeth with composite resin and replacement of the resorbed incisor with an acrylic denture. However, Case 2 was lost to follow up and no details of treatment can therefore be provided.

The authors postulate that the external root resorption seen in both cases could be a previously unreported part of the syndrome. They explore the possibility that the cementoblasts, or the cementum produced, are defective, resulting in exposure of dentine and subsequent root resorption.

As in all rare syndromes, many of the papers published on focal dermal hypoplasia comprise single case reports, each adding to the list of features. This paper, however, presents two such cases with external root resorption in addition to previously reported dental findings. It is hoped that this will aid in the diagnosis and management of such cases. Careful radiographic examination of patients with focal dermal hypoplasia should be carried out in order that any root resorption may be diagnosed and managed early.

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