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CASE REPORT

Histopathology of the teeth in segmental odontomaxillary dysplasia: new findings

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Histological examination of the deciduous teeth in two cases of segmental odontomaxillary dysplasia (SOMD) showed fibrous enlargement of the pulps, an irregular pulp/dentine interface displaying many pseudoinclusions and pulp stones. There were tubular defects in the coronal dentine from pulp horn to cusp tip, an irregular tubular structure to the circumpulpal dentine of the apical half, a focally deficient odontoblast layer and widespread external resorption. Together with the clinical features of unilateral maxillary enlargement, upper alveolar expansion in the distal segment, increased spacing and delayed eruption of the deciduous molars and absence of premolar teeth, these histological appearances allow distinction of this condition from fibrous dysplasia (FD), segmental hemifacial hypertrophy (SHH) and regional odontodysplasia (ROD).
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Introduction

Two 3.5-year-old boys were referred with symptomless persistent facial asymmetry, unilateral maxillary enlargement and unerupted upper deciduous molar teeth. In addition, there was expansion of the maxillary alveolus from the deciduous canine region posteriorly with thickening of the buccal gingiva. The deciduous canine and molar teeth were widely spaced and larger than normal (Fig. 1), with an ill-defined radiodensity of the affected maxillary alveolus (Fig. 2). Although these are the classical features of segmental odontomaxillary dysplasia (SOMD), in each case, neurofibromatosis and fibrous dysplasia (FD) were suspected. Gingiva and alveolar bone were biopsied; the deciduous canine tooth was removed in each case, along with the first deciduous molar tooth in Case 1.

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The histological changes in all the teeth were essentially similar. There were tubular defects in the coronal dentine extending from the pulp horn towards the cusp tip (Fig. 3). Most of the dentine in the crown and root was normal in appearance, but the circumpulpal dentine from the middle one-third of the root apically had an irregular tubular structure and lacked a well-defined odontoblast layer (Fig. 4). The pulp chamber had an irregular outline giving the impression of intradentinal pulpal inclusions with many pulp stones (Fig. 4). The pulp chamber and root canals were enlarged; the pulpal tissues were coarsely fibrous, vascular and moderately cellular with a distinctive whorled pattern (Fig. 4). Areas of external resorption were present on the apical aspects of the roots, some exhibiting partial repair. Immunohistochemical staining for S100 protein failed to demonstrate significant neural proliferation in teeth, bone or gingival tissues, helping to exclude neurofibromatosis.

Comments

The clinical and radiological features of SOMD, alternatively known as Hemimaxillofacial Dysplasia, are well described (1–4) but the condition is probably under-recognised, being diagnosed instead as an atypical form of FD, segmental hemifacial hypertrophy (SHH) or regional

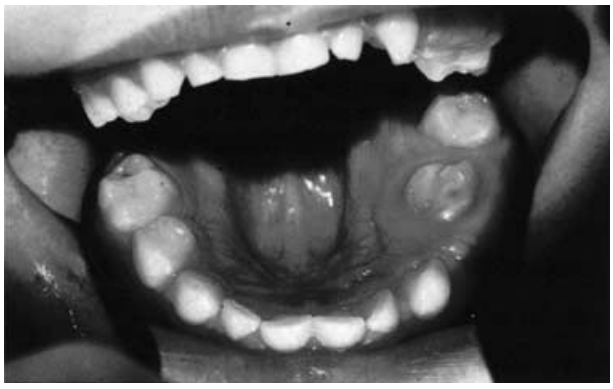

Figure 1 Case 1 – at presentation aged 3.5 years. Mirror view of expanded left maxillary alveolus with enlarged, spaced teeth and hypoplastic upper left deciduous first molar.

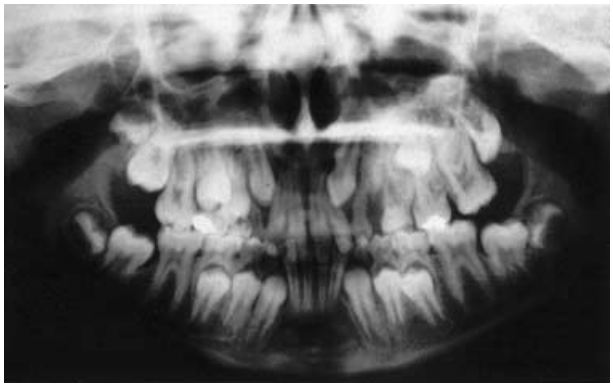

Figure 2 Case 2 – after 7.5 years follow-up aged 11 years. Diffuse ill-defined radiodensity in left maxillary alveolus. Note enlarged pulp chambers, the increased spacing of the deciduous and permanent molar teeth and the missing upper left first premolar.

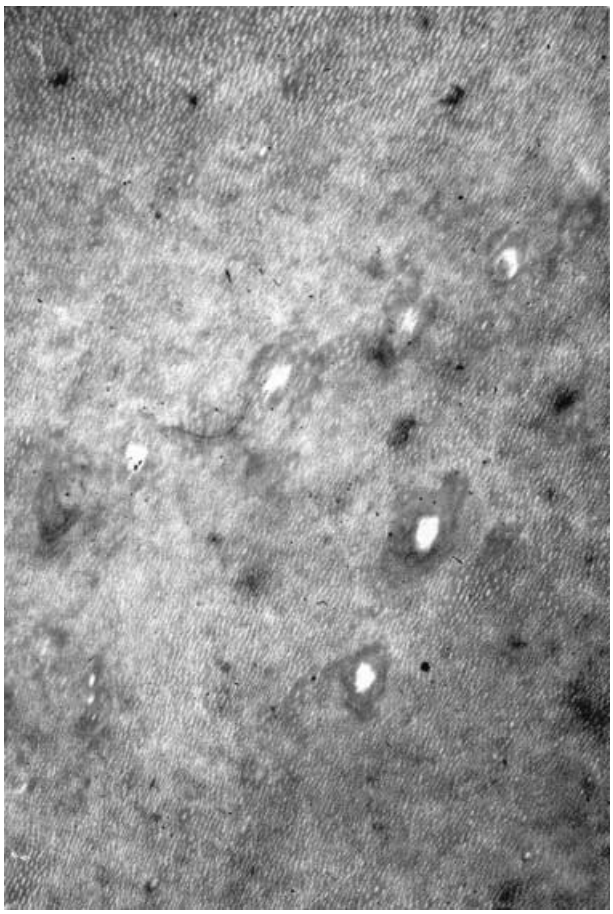

Figure 3 Case 1 – upper left deciduous first molar. Tubular dentine defects in coronal dentine (×400).

odontodysplasia (ROD). Histologically, affected gingival soft tissue is fibrous but uninflamed while the bone is immature with irregular coarse trabeculae (1, 4). Other dental abnormalities are recorded, most frequently missing premolar teeth and delayed eruption of permanent molar teeth in the affected segment (1–4), but this is the first account of the histopathological features of the teeth.

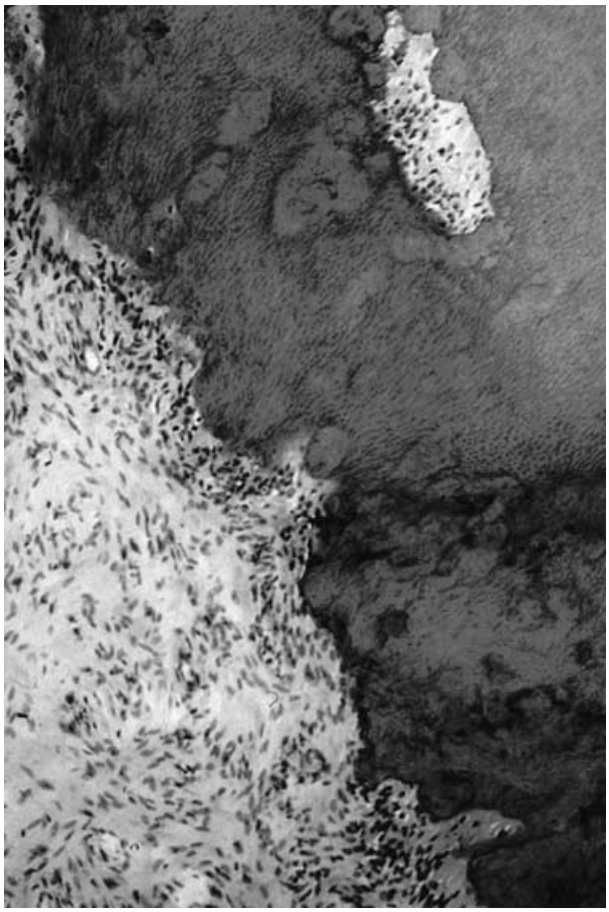

Figure 4 Case 1 – upper left deciduous first molar. Abnormal tubular structure of circumpulpal dentine, irregular pulp/dentine interface with pseudoinclusions, focally deficient odontoblast layer and coarsely fibrous pulp (×250).

SOMD can be distinguished from FD, SHH and ROD by correlation of clinical, radiological and pathological findings. The presence of dental malformations and soft tissue expansion of the gingiva excludes FD; in addition, there is much less tendency for progressive enlargement with SOMD. Although enlargement of the teeth may be seen in SHH, it is usually associated with an increased rate of shedding and replacement by the permanent successors. The unilateral enlargement in SHH affects other structures as well as the oral tissues but does not exhibit focal bony changes. Dental abnormalities and gingival thickening are features of ROD, occasionally accompanied by abnormalities of facial symmetry, but the radiographic and histological appearances of the 'ghost teeth' are characteristic; ROD is not restricted to the posterior maxillary segments and may even cross the midline (5).

The aetiology and pathogenesis of SOMD are not known but it is hypothesised to represent either an acquired developmental abnormality related to events in intrauterine life (1, 3) or to a post-zygotic mutation in a clone of progenitor cells, influencing further development in this part of the maxilla (2). We feel unable to contribute to this aspect of the debate but consider it unlikely that the dental changes are because of a primary defect in odontogenesis.

In conclusion, there are significant morphological defects in the teeth in our cases of SOMD, consideration of which, we believe, should facilitate recognition and diagnosis of further cases of this unusual condition.

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