

CASE REPORT

Segmental odontomaxillary dysplasia: clinical, radiological and histological aspects of four cases

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Segmental odontomaxillary dysplasia (SOD) is a rare developmental disorder of the maxilla, primarily involving the posterior part of the maxilla. Clinically, the disorder is often diagnosed in early childhood due to a unilateral buccolingual expansion of the posterior alveolar process, gingival enlargement, absence of one or both premolars in the affected region, delayed eruption of the adjacent teeth and malformations of the primary molars. In this report, four patients with SOD are described. The findings were similar to earlier reports, but for the first time an ipsilateral rough erythema on the skin in two of the subjects is reported.

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Hemimaxillofacial dysplasia (HMD) was first described in 1987 by Miles, Lovas and Cohen (1987). The disorder is characterized by facial asymmetry, facial hypertrichosis, unilateral maxillary dysplasia and hypoplastic teeth. In 1990, an additional eight patients with similar features were described (Danforth *et al*, 1990). Unlike the earlier report, Danforth *et al* did not report facial hypertrichosis. Because of the absence of facial features, Danforth *et al* suggested the term segmental odontomaxillary dysplasia (SOD). Like HMD, SOD is a rare developmental disorder of the maxilla, primarily involving the posterior part of the right or left maxillary bone, gingivae and dentition. Clinically the disorder is often diagnosed in early childhood due to a buccolingual expansion of the posterior alveolar process, gingival enlargement, absence of one or both premolars in the affected

region, delayed eruption of the adjacent teeth and malformations of the primary molars. Radiologically, the bone has been described as dense and sclerotic, and the histological findings have demonstrated immature bone with irregular trabeculae of woven bone with resting or reversal lines without osteoblastic rimming (Danforth *et al*, 1990).

A total of 27 cases with SOD/HMD have been reported (Prusack *et al*, 2000), but neither the cause nor the pathogenesis is understood.

We here report the progression of four new cases with SOD, two of which presented with an ipsilateral erythema of the skin not described previously.

Case reports

Four patients were referred to the Copenhagen School of Dentistry for consultation. The clinical, radiographical and histological features of the four subjects are given in Table 1.

Figures 1–4 show the first and last panoramic radiograph from subjects 1, 2 and 3; from subject 4 only one radiograph was available. Study casts from the first three subjects can be seen in Figure 5.

Clinical manifestations

Unilateral left maxillary enlargement was seen in all four patients, extending from the canine region to the tuberosity. Enlargement of both the buccal and palatal part of the alveolar process was noted, most pronounced on the buccal side. Abnormal spacing was observed between erupted primary molars and adjacent teeth. In subjects 3 and 4 surgical exposure revealed that the first molars were displaced buccally. A depression was observed in the midline of the palate in subject 1. In the canine region in all subjects a well-defined border was seen between the affected region and the adjacent normally developed region. In subjects 1 and 2 the canine erupted into a normal position and normal development and eruption of the left upper canine in subject 3 is also expected. Hypertrichosis was not present in any of the subjects, but a rough

Table 1 Clinical, histological and radiological appearance in four patients with SOD

	<i>Subjects</i>			
	<i>1</i>	<i>2</i>	<i>3</i>	<i>4</i>
Gender	Female	Male	Male	Male
Age at first examination (years)	3	8	2	3
Maxillary location	Left alveolar process First premolar to second molar	Left alveolar process First premolar to second molar	Left alveolar process First premolar to first molar	Left alveolar process First primary molar to tuberosity
Investigation period (years)	11–16	8–14	2–12	3–6
Skin manifestations	Erythema left cheek	No	Erythema left cheek	No
Maxillary alveolar enlargement	Yes	Yes	Yes	Yes
Gingiva	Enlarged	Enlarged	Enlarged	Enlarged
Bone radiographic appearance	Histology: slight fibrosis Sclerotic	Sclerotic	Sclerotic	Sclerotic
Bone histological appearance	Irregular bone structure indicating alternating resorption and formation of bone. Involucrum formation. Marrow spaces occupied by poorly cellular fibrous tissue	No histology	As in subject 1, but hypercellular bone without involucrum formation	No histology
Tooth morphology (in the affected region)	Splayed roots and obliterated pulp chambers of first and second primary molar. Atypical root resorption of primary molars	Splayed roots and progressive obliteration of pulp chambers of first and second primary molar. Atypical root resorption of primary molars	Splayed roots and progressive obliteration of pulp chambers of first and second primary molar. Atypical root resorption of primary molars	Splayed roots and large pulp cavities of first and second primary molar
Eruption (in the affected region)	Delayed: first premolar, permanent first and second molar	Delayed: first premolar, permanent first and second molar	Delayed: Permanent first and second molar	
Retained teeth (in the affected region)	Primary retention of second molar	–	–	–
Missing teeth (in the affected region)	First premolar and third molar	Second premolar	First and second premolar	Malformed or absent first premolar
Progressive growth	No	No	No	Too early to determine
Others	Panoramic radiograph shows minor involvement of the maxillary sinus	CT-scan: Lesion occupies most of the left maxillary sinus. No pathological activity is seen on bone scintigraphy	Panoramic radiograph shows no apparent involvement of the maxillary sinus	Panoramic radiograph shows minor involvement of the maxillary sinus
Treatment	One 3.3 mm ITI solid implant was placed in the affected region. Normal healing and osseointegration was observed. Observation period is 6 months.	Extraction of the primary molars in the affected region	First upper left molar was extracted	No treatment offered

erythema was observed on the skin on the ipsilateral side in subjects 1 and 3 (Figure 6). Furthermore, the mother of patient 3 stated that her son had been seen by a dermatologist and a diagnosis of childhood eczema was given for the erythema. The mother also reported that she had used many ointments on her son's face with no improvement.

Radiographical manifestations based on panoramic radiographs

Bone

Radiographical features of the patients were consistent and distinctive, showing sclerotic bone with thick and coarse trabeculae and poorly defined borders. Involvement of the ipsilateral maxillary sinus was noted in subjects 1, 2 and 4 (Figures 1a, 2b and 4).

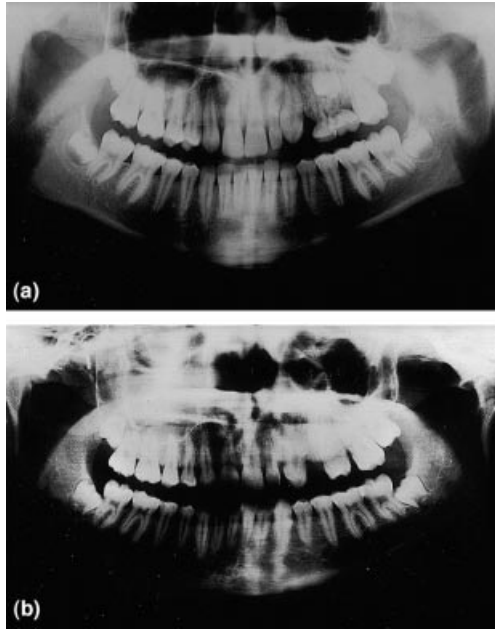


Figure 1 (a, b) Subject 1. Panoramic radiograph at age 11 and 16 years

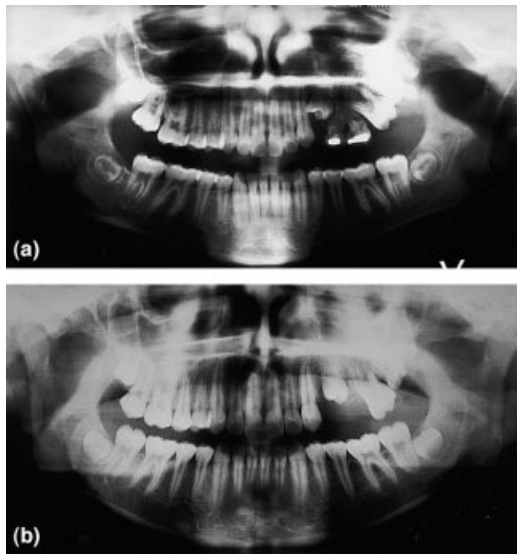


Figure 2 (a, b) Subject 2. Panoramic radiograph at age 8 and 14 years

Permanent dentition

Both premolars in the affected region were missing in subject 3 (Figure 3a, b), and one premolar was missing in subjects 1 and 2 (Figures 1a, b and 2a, b). The development of the first premolar in subject 4 was difficult to determine. In subjects 1 and 3 the first molars had a distal inclination (Figures 1a and 3b).

Primary dentition

Splayed roots, atypical root resorption and progressive pulpal obliteration were observed on most of the

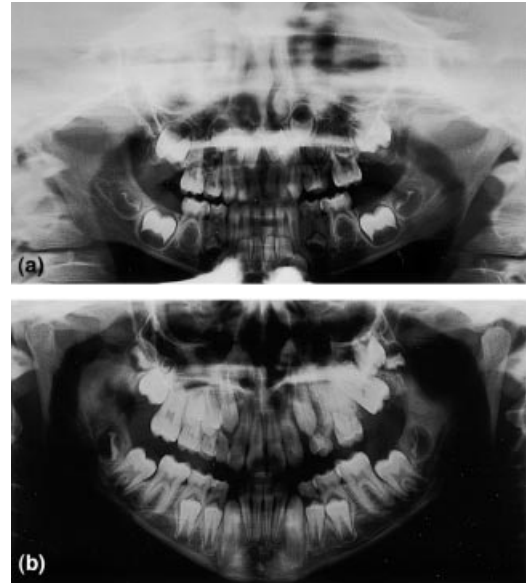


Figure 3 (a, b) Subject 3. Panoramic radiograph at age 10 and 12 years

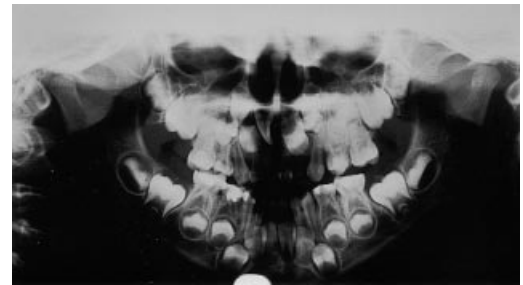


Figure 4 Subject 4. Panoramic radiograph at age 6 years

primary molars in the affected region (Table 1) (Figure 3a, b).

Histological manifestations

Case 1

Tissue from the gingivae covering a retained second molar revealed slight fibrosis but apart from this no pathological change were found. A biopsy from the bone in the canine region showed broad bone trabeculae with irregular basophilic cemental lines resulting from alternating resorption and formation of bone. The marrow spaces were occupied by poorly cellular fibrous tissue without inflammation and containing only a few fat cells (Figure 7).

Case 3

A biopsy from the bone in the region of the primary molars revealed similar features as described above. Furthermore, active bone resorption in some areas and distinct osteoblastic rimming in others were seen. The marrow spaces were occupied by a hypervascular, poorly cellular fibrous tissue without inflammation and

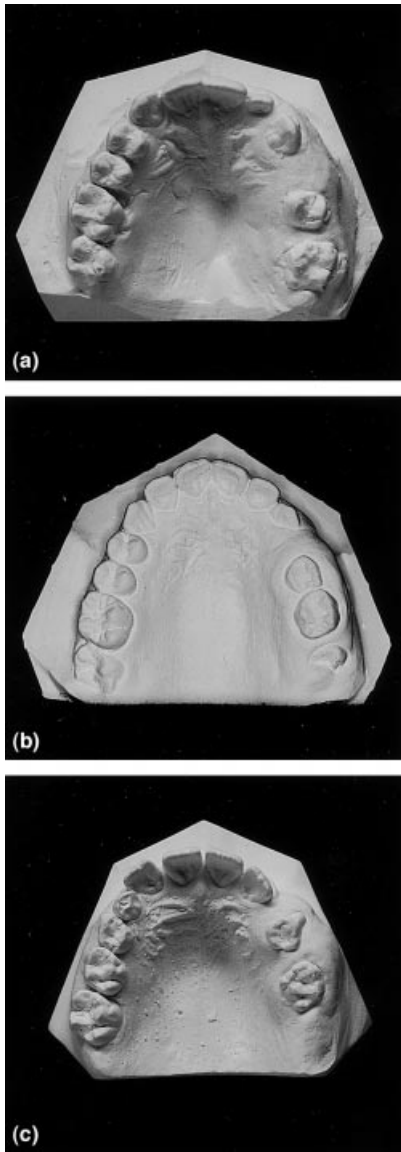


Figure 5 (a–c) Study casts from Subject 1 (a), Subject 2 (b), Subject 3 (c). Note similar enlargement of the alveolar process extending from the canine region to the tuberosity

containing only some fat cells. In one small area a rather cell-rich fibrous tissue was seen. A separate tissue fragment revealed a non-inflamed cystic lesion compatible with a dentigerous cyst.

Discussion

The four subjects described in this study have all the features of SOD, i.e. unilateral maxillary enlargement, missing premolars, malformations and atypical resorptions of primary molars and retarded eruption of adjacent molars (Danforth *et al*, 1990; Packota, Pharoah and Petrikowski, 1996; Prusack *et al*, 2000). The medical histories of the four patients were non-contributory. In all four patients the left side of the maxilla were involved which is probably a coincidence, as other cases of SOD



Figure 6 Rough erythema on left cheek in subject 1

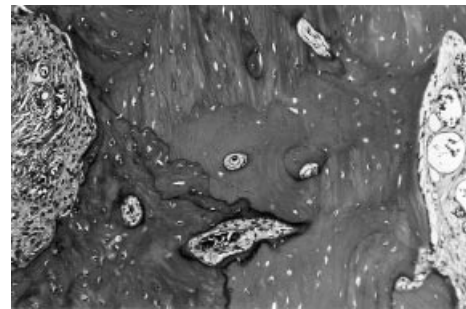


Figure 7 Photomicrograph showing compact bone with numerous basophilic reversal lines. Note bone resorption (left) and osteoblastic rimming right. Van Gieson. Original magnification $\times 125$

have been described involving the right side (Danforth *et al*, 1990; Packota *et al*, 1996).

Radiographically, thick and coarse irregularly oriented bony trabeculae producing a sclerotic appearance were observed in our patients in accordance with earlier studies (Danforth *et al*, 1990; Packota *et al*, 1996). A vertical orientation of trabeculae has been observed in some patients (Packota *et al*, 1996), which was not obvious in our patients. It was not possible to determine if the changes in the maxilla extended into the zygomatic bone.

A hitherto unreported finding was a rough erythema of the skin of the ipsilateral side in two of our patients. Hypertrichosis has been described in patients with HMD (Miles *et al*, 1987), and a hypopigmented area was observed in a patient with SOD on the same side as the maxillary abnormalities (DeSalvo *et al*, 1996). The erythema and hypopigmentation might have the same etiology as hypertrichosis expressed as a mild degree of skin involvement. A systemic or endocrine aberration was suggested by DeSalvo *et al*.

Differential diagnosis

Hemimaxillofacial dysplasia has similar manifestations as SOD, but includes facial hypertrichosis which is not seen in SOD (Danforth *et al*, 1990). It seems probable that the two diagnoses are different manifestations of the same disease. Interestingly, two of our four patients presented with erythema of the ipsilateral skin, further suggested a link between the two entities, basically separated by the presence or absence of skin abnormalities.

Regional odontodysplasia is characterized by delayed eruption, or failure of eruption of discoloured and atypically shaped teeth, with rough coronal surfaces that may be soft on probing. The condition appears, like SOD, to occur more frequently in the maxilla, involving the primary dentition equally, but the incisors and canines of the permanent dentition are more commonly involved. Missing premolars are not a typical feature of regional odontodysplasia (Crawford and Aldred, 1989). Radiographically, the affected teeth show reduced radiodensity (described as ghost teeth) because of the reduced thickness of hard tissues the adjacent bone does not exhibit structural changes. Histologically the dental changes are multiple and the enamel is irregular (Crawford and Aldred, 1989). Furthermore, typical calcifications are seen in the dental follicles. Clinically, radiographically and histologically, regional odontodysplasia can therefore be distinguished from SOD.

Microscopically, SOD does not display the typical features of fibrous dysplasia in which irregularly shaped individual bony trabeculae are formed in a rather cellular, loosely arranged fibrous stroma. In mature fibrous dysplasia of the jaws larger trabeculae of lamellar bone often arranged in a parallel pattern in a moderately cellular fibrous stroma can be seen. However, in the cases reported here the bone did not assume the form of irregular trabeculae but rather represented changes as a result of alternating resorption and formation of bone. Bone affected by fibrous dysplasia grows out of proportion to unaffected bones, producing enlargements and deformities, and tooth malformations and missing premolars are not characteristic of the disease (Zimmerman, Dahlin and Stafne, 1958). The bony involvement in SOD seems to be not progressive at an early stage of life as judged from our cases and the literature.

In hemifacial hyperplasia the involved area may vary from a single digit, a single limb or unilateral facial enlargement. Oral manifestations include precocious eruption in the affected region and enlarged teeth. Absence of premolars has not been reported (Gorlin, Cohen and Levin, 1990).

The initial event of the SOD condition is unknown. The early manifestations in some subjects and absence of one or both premolars suggests that the disturbance probably occurs *in utero*, at birth or in early infancy. A localized viral infection or localized tissue ischemia have been suggested as intrinsic causative factors in regional odontodysplasia (Buyse, 1990). Because of the very

consistent distribution of SOD it could be speculated that a viral or bacterial infection along the branches of the maxillary division of the trigeminal nerve could be the initial causative factor.

The eruption of the permanent teeth in the affected region is retarded compared with the contralateral side. Emergence through the gingivae is possible, but it seems that the premolars and molars are partially retained (secondary retention). Obviously a resorption path through the dysplastic bone can be established, but the craniofacial growth exceeds the eruption speed/capacity and the teeth do not come into occlusion. Normally, secondary retained teeth are ankylosed, but this is seemingly not the case in these patients with SOD.

A total of 31 cases of SOD/HMD have been reported to date including this report, but it is believed that many cases have been diagnosed as fibrous dysplasia (Zimmerman *et al*, 1958; Packota *et al*, 1996). Therefore the incidence of SOD is unclear.

Many reported cases of SOD have been directed primarily to oral and maxillofacial surgeons. This report attempts to increase the knowledge about SOD among general dentists, and furthermore to encourage long-term treatment and follow-up reports which are lacking in the literature.

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