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CASE REPORT

Dental findings and rehabilitation in familial osteodysplasia (Anderson type): a case report

S Deger[1], G Tabar[1], B Sermet[1], H Tanyeri[2], E Kurklu[2]

Departments of [1]Fixed Prosthodontics, and [2]Oral Surgery and Oral Medicine, Faculty of Dentistry, Istanbul University, Istanbul, Turkey

Familial osteodysplasia is a disorder of osteogenesis with an autosomal recessive pattern of inheritance which predominantly affects facial bones. No recent case had been reported, particularly from a dental point of view since the syndrome was first described by Anderson *et al* (*JAMA* 1972;220:1687–93). A 23-year-old male with familial osteodysplasia was presented in maxillofacial and dental aspects with clinical and radiological manifestations including malocclusion, abnormal teeth alignment, impacted teeth, shape disturbances including uncompleted coronal formation, root shortening with bulbous form, high angled mandible and elongation of the corpus of mandible. Recognation of the syndromal features prior to any dental intervention is of paramount importance because of increased inclination to spontaneous mandibular fractures. Hence, no surgical intervention was performed for impacted teeth. Following the extractions of severely mobile teeth, a definitive restoration was fabricated as distal-extension removable partial dentures with conus crown telescopic system. The aesthetic and functional outcome was satisfactory for the patient. In conclusion, dentists appear to play an important role in the recognition of familial osteodysplasia, based on maxillofacial and dentoalveolar findings. Awareness of the syndromal features, especially of spontaneous fractures, would detect the limitations for dental interventions and treatment planning.
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Keywords: familial osteodysplasia; conus crown; mandibular fracture

Introduction

Familial osteodysplasia was first defined by Anderson *et al* (1972) as ' a new syndrome involving distinctive abnormalities of the facial bones'. The most distinctive abnormalities involve facial bone, calvarium, spine, clavicles, ribs, pelvis, femur and feet. The craniofacial defects consist of midfacial hypoplasia, flat nasal bridge, pointed chin, depressed zygomatic bones, hypoplasia of the petrous bone, calvarial thinning, prominent eyebrows, large ear bones, mandibular prognathism, mandible with wide angle, micromaxilla, recurrent mandibular fractures and malocclusion. Diastolic hypertension and hyperuricaemia also appear as common clinical findings (Anderson *et al*, 1972; Buchignani *et al*, 1972).

The syndrome has an apparent autosomal recessive pattern of inheritance (Anderson *et al*, 1972; Buchignani *et al*, 1972). It was defined in detail with genetic, radiologic and physical properties, but no recent cases have been reported since 1972 when the syndrome was first described. Also there exists no dental literature related to clinical manifestations of dental formation, shape abnormality, distribution and dental treatment. Only a report of Shendel and Delaire (1982) that mentioned craniofacial features of the syndrome together with partial dental agenesis was presented.

As the syndrome primarily affects the maxillofacial and alveolar bones, dentists possibly appear to be the first to detect the abnormality and make a diagnosis. The increased inclination to mandibular fractures plays an important role in the treatment choice. Refraining from excess force during dental treatment and providing optimal occlusal stress distribution in the prosthetic planning are essential in familial osteodysplasia.

The aim of the report was to draw attention to this rare and lesser-known syndrome and to present the maxillofacial and dentoalveolar manifestations along with the limitations for dental interventions and options of treatment which are lacking in the dental literature.

Case report

A 23-year-old male patient attended the clinic with an expectation of having implant-supported fixed prosthesis for the provision of his aesthetic and functional demands. The patient was a healthy looking man with a

Correspondence: Sabire Deger, İstanbul Üniversitesi, Dişhekimliği Fakültesi, Protetik Tedavi A.B.D., Kuron-Köprü Protezi B.D., 34393, Çapa, İstanbul, Turkiye. Tel: +90 (0) 212 414 202 20/30295, Fax: +90 (0) 212 531 22 30, E-mail: sabdeger@yahoo.com
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V-shaped facial appearance presenting a pointed chin, thick and bushy eyebrows, large ear bones and a long and bulbous nose (Figure 1a,b).

The intraoral examination revealed malocclusion, abnormal alignment, impacted teeth, and shape disturbances with a poor oral hygiene. Shape disturbances consisted of slightly ambiguous clinical morphology resembling primary teeth and notched enamel at the tips of the cusps and incisal margins displaying the colour of dentine. The teeth 15, 27, 33 and 37 had no crowns, but only retained roots which were shortened and severely mobile. The molar teeth except for tooth 26 displayed gingival recession up to furcation. No soft tissue pathosis was observed.

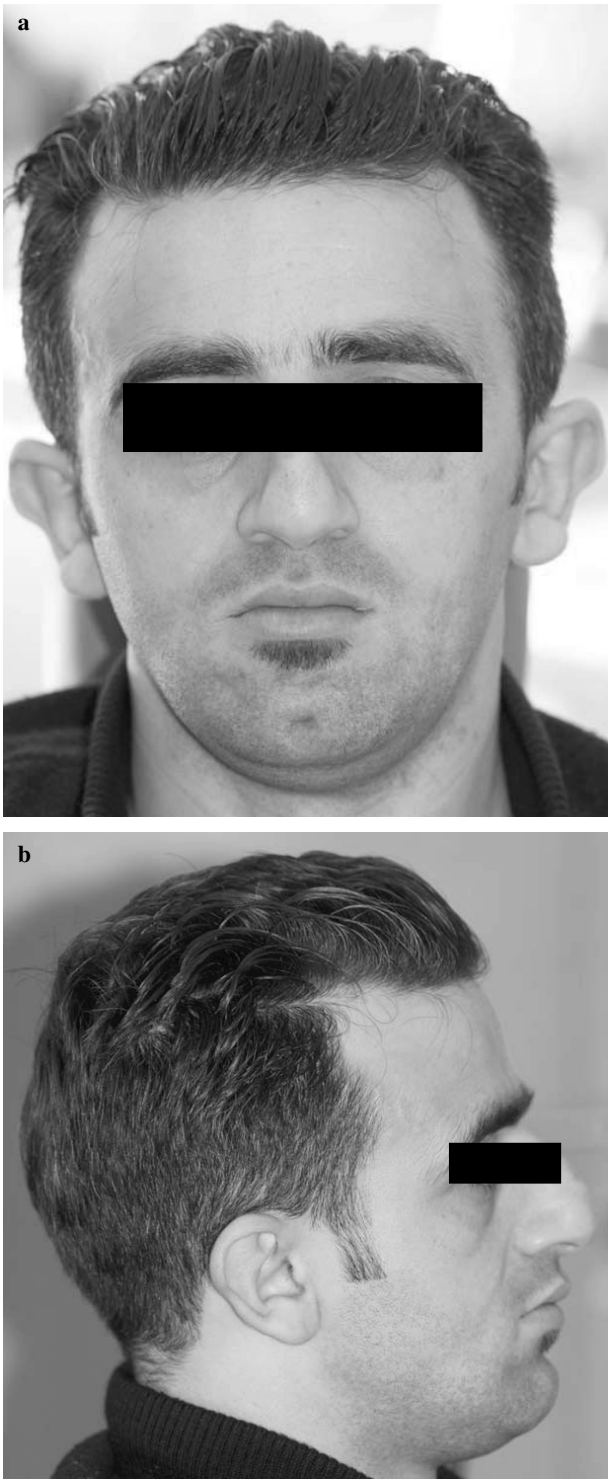

Figure 1 Facial appearance of the patient. Peculiar facial features are obvious. (**a**) Frontal aspect. (**b**) Profile aspect

Panoramic view revealed the impaction of teeth 13, 14, 18, 28, 35, 38, 45 and 48 with uncompleted root formation with a bulbous morphology. A resorptive process affecting the supporting alveolar bone around all remaining natural teeth was detected (Figure 2). Lateral cephalometric analysis revealed a high angled mandible (NSGn: 78°, S-N/Go-Gn: 45°), elongation of the corpus of mandible (Go-Me: 75 mm), protrusion in the upper incisors (U1/NA: 5 mm/35°, U1/S-N: 108°) and retrusion in the lower incisors (L1/NB: 3.5 mm/13°, L1/Go-Me: 77°) (Figure 3).

Developmental and eruptive abnormalities of the teeth together with facial appearance and remarkable family history of consanguineous marriage of parents had led to a doubtful genetic cause. The patient was then referred to the Division of Medical Genetics of Istanbul University for further investigation. Radiologic

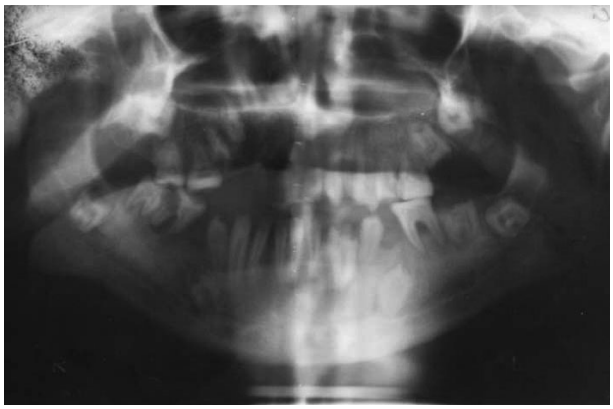

Figure 2 Initial panoramic view

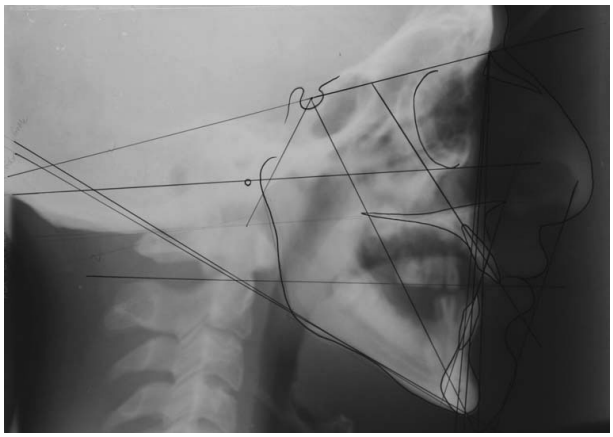

Figure 3 Lateral cephalometric analysis

examination, haemotologic and urine analysis, bone density measurement and clinical assessment revealed the diagnosis of familial osteodysplasia syndrome of Anderson type.

As spontaneous bone fractures are peculiar features of this syndrome, no surgical intervention which may lead to any bone loss was performed for the management of impacted teeth. Extractions of the teeth 15–17, 33, 36, 37, 46 and 47 were performed because of severe mobility (Figure 4). The maxillary left second premolar and the first molar teeth were subjected to endodontic therapy because of extensive decay and periapical lesion. Although teeth 24, 25 and 34 displayed a crown/root ratio of 1:1 and mild mobility, they were restored with respect to their key role in the maintenance of centric occlusion and vertical dimension (Figure 5).

The patient was young and had high aesthetic demands. Implant-supported fixed prosthesis, as the patient requested, could have been an option but this was out of question because of the increased inclination to spontaneous bone fractures. Hence, a treatment option including conus crown telescopic system was chosen. There was contact between the right tuber region and the retromolar region at centric occlusion, which was a boundary for any prosthetic construction (Figure 5). Alveoloplasty was performed at the right

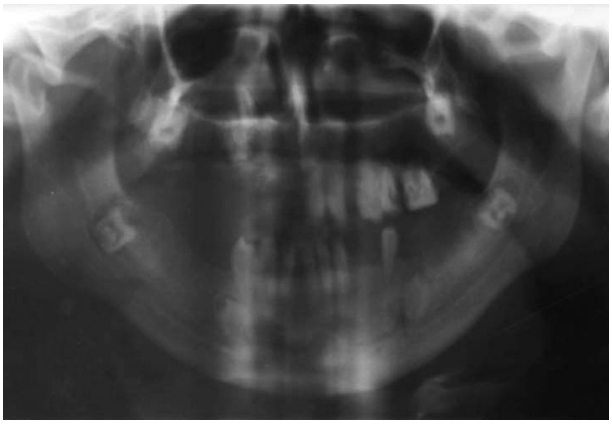

Figure 4 Panoramic view after tooth extractions

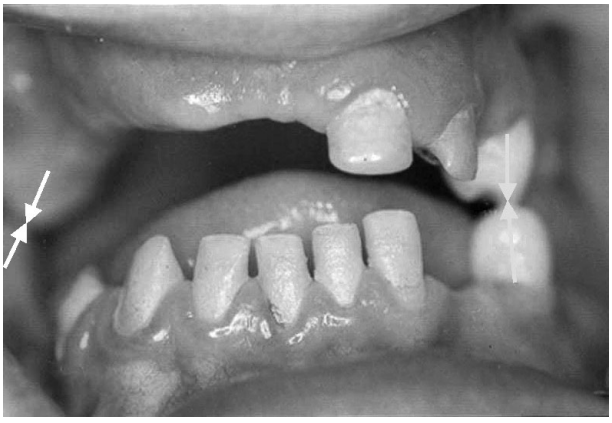

Figure 5 Vertical stop between teeth 25 and 34 at the left (green arrow) and the contact between right tuber region and retromolar region (white arrow) are seen

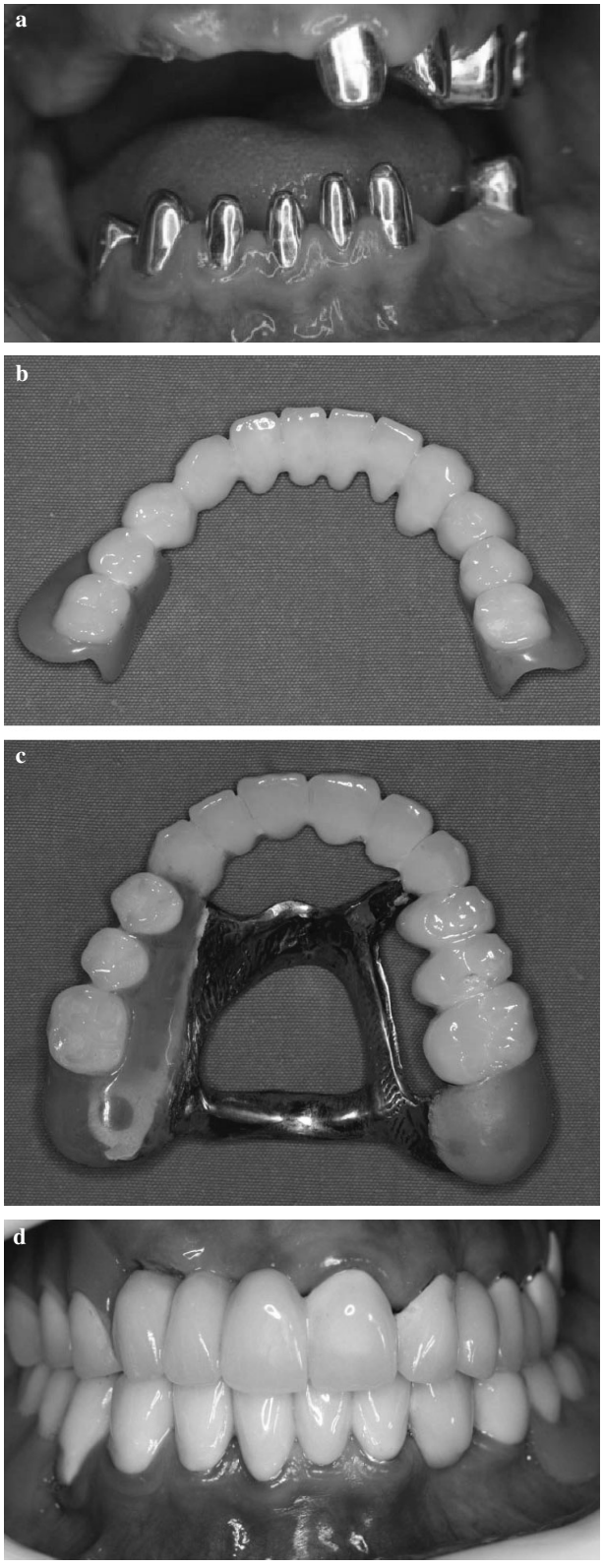

Figure 6 (**a**) Inner crowns in place. (**b**) Distal-extension removable lower partial dentures. (**c**) Distal-extension removable upper partial dentures. (**d**) Definitive prostheses are placed

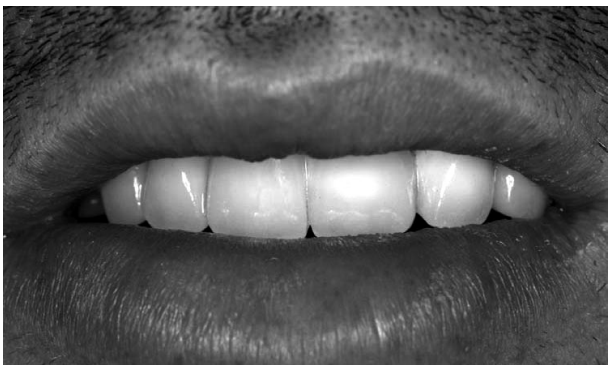

Figure 7 View of the prostheses at smile

tuber region under local anaesthesia. The bone sample was histopathologically examined and no pathology, but new thick bone formation, was detected. Centric relation was accomplished by the vertical stop at the contact between teeth 25 and 34 because of the infraocclusal position of the molars. Initial and temporary removable acrylic resin dentures were constructed to provide a functional occlusal relationship over a period of 3 months. Definitive restoration was fabricated as distal-extension removable partial dentures with conus crown telescopic system (Figure 6a–d).

The aesthetic and functional outcome was satisfactory for the patient (Figures 6d and 7). During the fabrication period of definitive restoration of tooth 13 which was impacted initially started to erupt and the incisal margin of the tooth appeared in a pointed form and with an uncompleted enamel structure. Functional loading is believed to activate the eruption and surgical extraction was not performed in order to refrain bone loss. The tip of tooth 13 was ground and as the eruption continued the palatinal surface of the prosthesis was ground to provide sufficient space for the tooth but the eruption eventually ceased. A follow-up period of 3 years was uneventful. Stabilization of the abutment teeth was maintained as the postrestorative twelfth month.

Discussion

Familial osteodysplasia is a disorder of osteogenesis which predominantly affects facial bones with an autosomal recessive pattern of inheritance. The possibility of heterogeneous parents to have an homozygous child is one in four. In the current case, while the parents and the siblings were not affected, the proband is believed to be a homozygous. A possible genetic disturbance, occurring during an unknown stage of osteogenesis and bone maturation, is thought to be the causal factor for the disorder. In comparison with other facial deformities of genetic origin, the anatomical deformations in familial osteodysplasia are related to bony structures, particularly the craniofacial skeletal structures. The patient reported here displayed characteristic craniofacial features of the syndrome, which actually were not discriminative for a clinician to suspect a syndrome or a disorder. Besides, some of the findings, when seen as an isolated instance, can be accepted as normal and some patients may be affected to a greater extent than the others. The patient was not aware of the syndrome he carried, before he attended the dental clinic. He had no symptoms, no history of fractures and had not had a detailed dental examination until he was 23 years of age. He did not suffer from any bothering symptom such as diastolic hypertension or hyperuricaemia. Biochemical analysis generally does not reveal diagnostic changes in patients with familial osteodysplasia. Neither did we detect any abnormality in blood and urine analysis.

Prognathism and malocclusion are common features of familial osteodysplasia. Shendel and Delaire (1982) reported a family with familial osteodysplasia presenting maxillo-mandibular abormalities leading to prognathism and partial dental agenesis. In this case, although first clinical assessment seemed to reveal partial agenesis, detailed panoromic view and intra-oral examination disclosed impaction of multiple teeth. Besides, remaining roots associated with uncompleted anatomical crown structure especially of the molars and diminution in the clinical crown/root ratio were also presented as dental manifestations.

Dental treatment may be damaging for patients with familial osteodysplasia because trauma associated with dental management may cause undesired consequences related to increased inclination to mandibular fractures. Anderson *et al* (1972) reported a woman with familial osteodysplasia presenting a history of mandibular fracture during an attempt for extraction of a right mandibular molar. Hence, recognition of the syndromal features prior to any dental intervention is of paramount importance. Prosthetic planning becomes a troubling issue in such cases because of malformation and abnormal lining of the teeth and also their questionable future as abutment teeth. The load distribution of prosthetic reconstruction is generally considered to depend on the design of the prosthesis. Optimal balance between the design of a prosthetic reconstruction and its capacity to withstand loading of the supported tissues is important for the long-term success of prosthetic therapy (Körber, 1983; Deger and Saadat, 1998b). In the present case, with the telescopic denture, the stress applied on the abutment teeth was distributed over a relatively wide area of the alveolar bone. Such a design also provided a secondary splinting effect on the supporting teeth. Besides this, the conus crown telescopic system allows removal of the superstructure when there is a need for additional periodontal or endodontic therapy and extraction of failed abutments after the completion of prosthetic treatment (Körber, 1983; Deger and Saadat, 1998a,b). The use of conventional telescopic prosthesis may not be recommended when there is a high aesthetic demand. Fortunately, the patient did not have a high lip line at smile or thin, delicate gingival tissue in the anterior region. Hence the aesthetic outcome was very satisfying owing to the metal-ceramic construction as well.

In conclusion, the dentist may play an important role in the recognition of familial osteodysplasia, based on maxillofacial and dentoalveolar findings, because this condition is often unrecognized by general physicians or because of the lack of or slightness of extracranial syndromal features. Consequently, this can have a major impact on the general health of the patient and also influence the planning of dental treatment.

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References

Anderson LG, Cook AJ, Coccaro PJ, Coro CJ, Bosma JF (1972). Familial osteodysplasia. *JAMA* **220:** 1687–1693.

Buchignani JS, Cook AJ, Anderson LG (1972). Roentgenographic findings in familial osteodyplasia. *Am J Roentgenol* **116:** 602–608.

Deger S, Saadat F (1998a). Hybrid prosthesis application with conical crown. *J Dent Fac Istanbul Univ* **32:** 10–15.

Deger S, Saadat F (1998b). Telescopic systems. *J Dent Fac Istanbul Univ* **32:** 71–76.

Körber K (1983). *Konuskronen. Das rationelle Teleskopesystem Einführung in Klinik und Technik*. Hüthig: Heidelberg, pp. 64–153.

Shendel SA, Delaire J (1982). Familial osteodysplasia. *Head Neck Surg* **4:** 335–343.