Case Report

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Idiopathic Juvenile Osteoporosis: A Case Report

Dentistry Section

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ABSTRACT

Idiopathic Juvenile Osteoporosis (IJO) is a very rare disease, self restrictive and shows marked, spontaneous improvement during adolescence. The major clinical features were pain with difficulty walking, growth retardation, oral and dental abnormalities with radiographically porous bone structure. A 13-year-old male referred to paediatric dentistry clinic for toothache. The observations made with extra-intraoral clinic examination that one revealed short and skinny stature, diffuse caries in deciduous teeth, abraded lower incisor, deep bite and dysmorphic appearance in permanent incisor. This report emphasizes the recognized features of IJO as well as describes facio-dental findings that could aid in the diagnosis and management of these patients.

Keywords: Bone, Diagnosis, Paediatric dentistry, Tooth

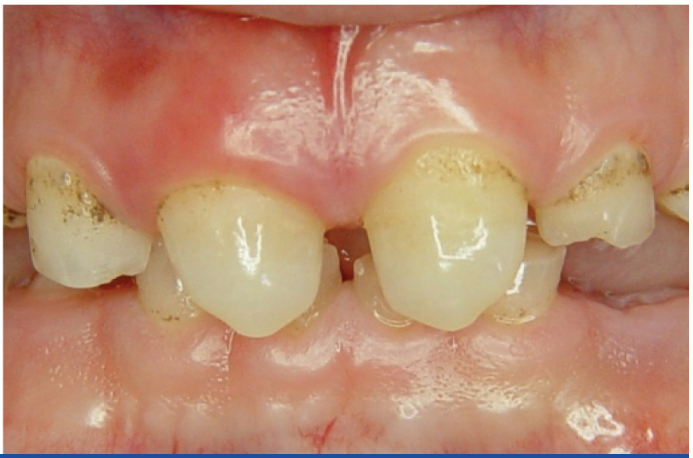
CASE REPORT

A 13-year-old male was referred to the Selcuk University's Faculty of Dentistry Department of Paediatric Dentistry Clinic, Konya, Turkey for a toothache in 2012. The clinical history revealed that he had also pain in his back, hips, and legs. There was no history of trauma, no history of chronic pain in back/extremities at family members or bone disease, eating disorder or systemic complaints. An intra-oral examination identified severe caries in 55,65; occlusal caries in 36, abraded 71,72,81,82 and a conical dysmorphic appearance of 11, 21 accompanied by deep bite [Table/Fig-1,2]. The absence of 11,14,15, 22-25, 31,32,35,41,42,45 and radiolucent areas in the periapical of 55,65 were noted during radiological examination [Table/Fig-3].

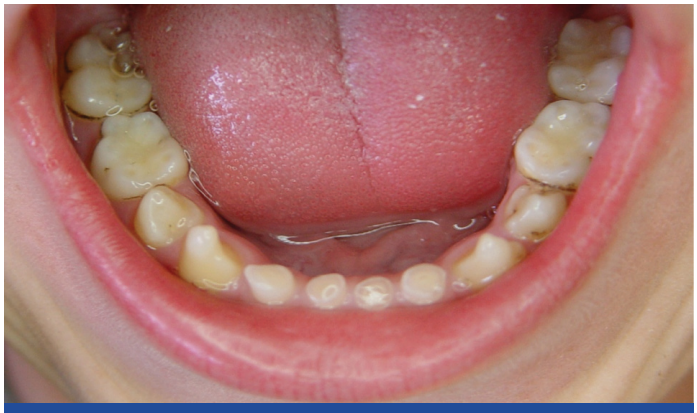
The patient was further evaluated clinically due to his markedly short stature and weakness [Table/Fig-4]. Findings included: a height of 136 cm (<3rd percentile), body weight of 30.5 kg (<3rd percentile), vital signs were within normal limits, normal appearance and signs of puberty concordant with age. Other systemic examination results were normal.

Laboratory examinations of his haemogram, serum glucose, hepatic and renal functions, celiac panel, acute phase reactants, thyroid (4.2 µIU/mL), parathyroid (53.23 pg/ml), growth hormone (3,43 ng/ml), Adrenocorticotropic hormone (ACTH) (29.95 pg/mL), calcium (4.6 mg/dL), phosphate (4.6 mg/dL), alkaline phosphatase (225 u/L), 25-hydroxyl vitamin D levels (20 ng/mL), urine and stool analyses and urinary calcium elimination (<2.0 mg/dL) were found to be within normal limits.

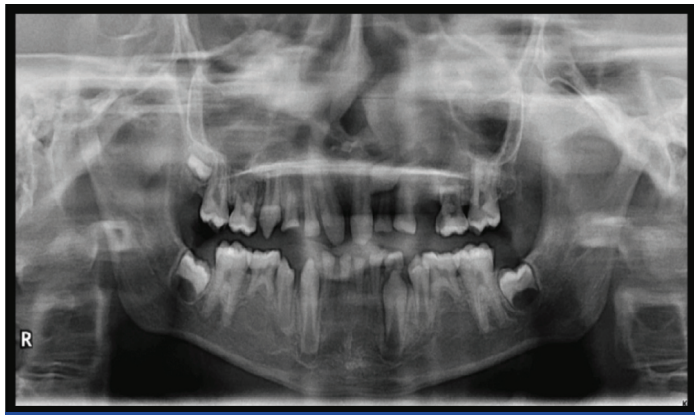
Bone mineral density (BMD) was 0.348 g/cm^2 (L1-L4) with a Z score of -4.1 (osteoporotic). The patient was diagnosed with IJO. In addition to the recommendations on nutrition and exercise, the patient was administered zinc (15 mg/day) (Zinc Nutrimed, Nutrifarma, Istanbul, Turkey), calcium 1000 mg (Calcium-Sandoz® Forte, Novartis, Germany) and vitamin D 1600 units/day (Devit 3 drops-Deva, Turkey).



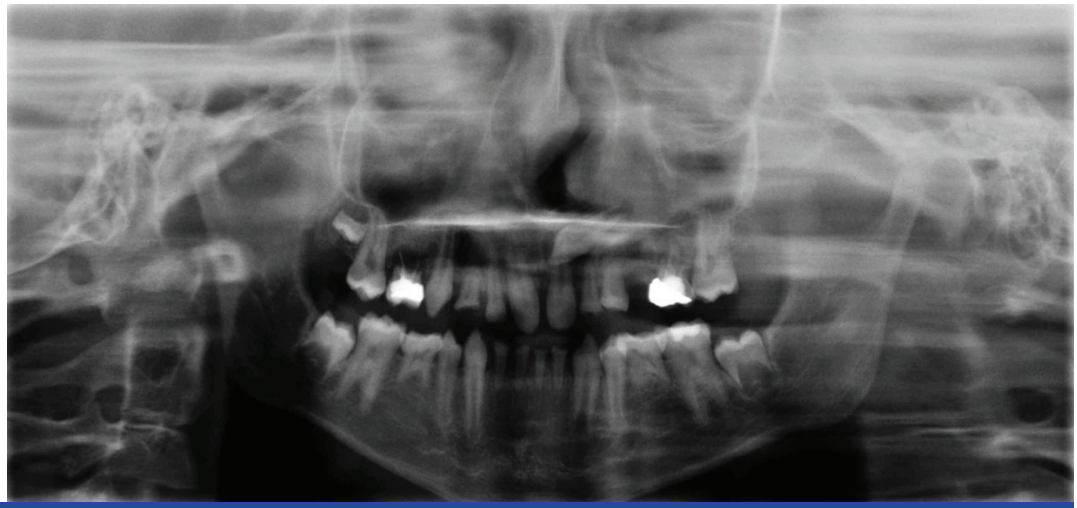
[Table/Fig-1]: Conical and dysmorphic permanent upper incisor and deep bite



[Table/Fig-2]: Lower abraded incisor



[Table/Fig-3]: Radiological examination before treatment



[Table/Fig-4]: Short stature and weakness appreance of patient **[Table/Fig-5]:** Radiological exmanination after treatment and periapical lesion on upper right side

Dental treatment was planned as a root canal treatment for deciduous upper second right and left molars. A 17% solution of Ethylene diaminetetraacetic acid (EDTA) (Sultan, Tehran, Iran) was used for the sclerotic channels during the canal preparation of the second upper molars. The canals were filled with guttapercha (Diadent, Burnaby, Canada) and root canal sealer (Sealapex, Romulus, US). Deciduous upper second right molar was restored with composite and deciduous upper second left molar with amalgam filler for the extension of the cavity borders beneath the gingival.

During the first year of patient monitoring; intraorally severe mobility, mild mobility was revealed in 65 and 55, respectively. Radiographically, a broad periapical lesion was identified in 65 with OPG, which were subject to extraction [Table/Fig-5]. Consequently, 65 and 55 were extracted and sent for pathological evaluation. The examination revealed diffuse resorptive activity associated with the root canal filling on the root site. The patient is on regular follow up by paediatric dentist. Because of having fragile bone structure, people with IJO are explained to perform slight training, prefer slight exercises and be careful about falling during playing or hard strikes. They are also advised to benefit from sun light and eat Ca supported foods. Informed consent form was signed by the patient for all medical and dental examinations, tests and treatments.

DISCUSSION

Osteoporosis is a complex, multi-factorial, systemic disease characterized by low bone mass and reduced bone resistance associated with micro-structural defects in the bone tissue, and an increased risk of bone tissue fracture [1].

Idiopathic juvenile osteoporosis (IJO) is a rare disease affecting children between 8 and 14 years of age. It is self restrictive and shows marked, spontaneous improvement during adolescence. The pathogenesis of the disease has not been completely established [2].

IJO is very difficult to diagnose. Patient history, physical examination and laboratory findings are considered, and the disease can only be diagnosed if other conditions such as osteogenesis imperfecta and secondary osteoporosis are excluded. Diseases of the connective tissue, Cushing's syndrome, Celiac disease, rickets, Wilson's disease, hyperthyroidism/hyperparathroidism inflammatory bowel disease, cystic fibrosis, thalassemia, chronic renal failure, cerebral palsy, hypophosphatasia, delayed puberty, and immobilization are among the causes of secondary osteoporosis [3].

Osteoporosis is diagnosed based on BMD level of < -2 score levels. Accordingly, a Z score of -2 or below means "lower bone density than expected for the chronologic age", and a Z score higher than -2 "normal bone density for the chronologic age". The patient's Z score was -4.1. In this case, we excluded rickets by normal values of serum phosphate, calcium, alkaline phosphatase and absence of characteristic radiological changes. Endocrinal diseases were ruled out by hormonal assays. Normal ranges of serum urea, creatinine and blood gases excluded renal and metabolic causes. The patient has a markedly short stature and low body weight signs that support typical symptoms of IJO [2]. Physical examination and laboratory findings excluded the causes of secondary osteoporosis while his age and symptoms of our patient supported IJO [4,5].

An osteogenesis imperfecta diagnosis had to be excluded as it can manifest dental signs and is known to produce symptoms similar to IJO. Physical examination however showed that colour of our patients sclera was white. He also had white deciduous and permanent teeth, lacked hypoplastic and/or hypocalcific defects of enamel and dentine and showed no evidence of hearing loss. Normal eye examination excluded osteoporosis pseudoglioma syndrome. Absence of dentinogenesis imperfecta, blue sclera, deafness excluded osteogenesis imperfecta types 1, 2, 3 and 4 [6]. While according to the literature, atypical crown formation was not thought to be characteristic of IJO, no adequate studies that would conclusively prove the absence of an association with IJO, were found [7].

In addition to third molars, the congenital deficiency of six or more teeth is called oligodontia or severe hypodontia [8]. The patient was diagnosed with oligodontia due to the deficiency of a large number of teeth on the panoramic radiograph. Intraoral signs in terms of maxillar incisors were notable for the view of an atypical crown. Oligodontia may also be associated with a certain number of syndromes [8]. Evaluating our patient in terms of tooth loss, oligodontia is often shown due to a decrease in the bone structure with increasing onward age [9,10]. Atypical crown morphology and congenital oligodontia might be considered dental findings of IJO.

Patients with IJO require a more conservative dental approach, as it is associated with a fragile bone structure [11]. The symmetric periapical dental lesions seen in 55 and 65 after one year, root canal treatment would suggest that it stimulates osteoclastic activity in the porous bone structure. The result of pathological examination also supports this opinion. Because of osteoclastic activity seen in present case, tooth extraction may be regarded primarily instead of root canal treatment in deciduous tooth with congenital absent permanent tooth.

The literature review revealed that there are limited studies regarding the intra-oral findings of IJO. Furthermore, to the best of our knowledge, there are no studies determining dental oral findings and dental treatment procedure, following osteoclastic activity in deciduous teeth in atypic incisor morphology with IJO. The reason for this may be the fact that the onset of IJO occurs during a limited period during pre-puberty and that bone structure

returns to normal after the period of pubertal development and with medical treatment. Intra-oral symptoms may not be noticed by the dentists during these short intervals. This, in turn, results in a lack of information among dentists, which then causes the diagnosis to be overlooked. Intra-oral findings of IJO should be presented in the literature through epidemiological studies.

CONCLUSION

Presentation of such cases will improve the knowledge of relation of IJO with facio-dental findings and dental managements. As a consequence, early diagnosis and the required medical treatment of cases of IJO will be possible, and porous bone structures will be reverted to their normal state within a short period of time.

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