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Case Report

A Rare Case of Multiple Dentigerous Cysts in Patient with Osteogenesis Imperfect a Type 1 A: Case Report

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ABSTRACT

Osteogenesis imperfecta is a rare inherited genetic disease characterized by increased bone fragility due to defective synthesis of the collagen molecule. Dentigerous cyst is the most common odontogenic developmental cyst associated with impacted teeth. Till date, in literature no association has been reported between the dentigerous cyst and osteogenesis imperfecta. Here in this case report, we present a rare case of osteogenesis imperfect a type 1A representing with multiple dentigerous cyst central type; the first case to be reported in the literature till date to the best of our knowledge.

Keywords: Osteogenesis imperfect, dentigerous cyst, Blue sclera, mandibular prognathism.

INTRODUCTION

Osteogenesis imperfecta also known as a brittle bone disease is the rare genetic disorder associated with improper collagen synthesis. Since its first description by Sir Olof Jakob Ekman in 1788 in his doctoral thesis as "osteomalacia congenital." [1] osteogenesis imperfecta is been identified by various other names as osteopsathyrosis idiopathica, Vroliks syndrome, Lobstein syndrome. [2] Sillence DO, et al classified the condition into four types based on clinical features and severity. [3] Though the most commonly identified gene for the condition is COLA1 and COLA2, advanced genetic research has led to the identification of multiple genes responsible for the condition and thus expanding the Silence classification into 15 other types. [4] Silence et al reported a prevalence of 3-4/100,000 and an incidence of 3.5/100,000 for OI type I osteogenesis imperfecta.

Dentigerous cyst is usually found in association with impacted teeth, known to occur due to the accumulation of fluid between dental follicle and tooth structure, or between epithelial layers of the lining of dental follicle.

Among the various facial and craniofacial abnormalities in patients of osteogenesis imperfecta, the association between dentinogenesis imperfecta and osteogenesis imperfecta is the widely reported phenomenon. Type I and Type IV osteogenesis imperfecta is subclassified as type A if dentinogenesis imperfect is absent and type B if dentinogenesis imperfect is present. Association between fibro osseous lesion and osteogenesis imperfecta is fairly reported in the literature; mixed radiopaque and radiolucent fibroosseous lesions mainly in the tooth bearing area of the jaw of 13 patients with osteogenesis imperfecta was reported by Levis LS et al. [5] A case of

cementifying fibroma in patient with osteogenesis imperfecta was reported by ShibaraT in 1996. [6]

A variant of osteogenesis imperfecta known as osteogenesis imperfectacystica, [7] known to represent with cystic lesions of the long bones in a patient with osteogenesis imperfecta has been reported in the literature.

Only 2 cases of osteolytic lesions in consistent with radiographic features of cyst in the jaw bones of a patient with osteogenesis imperfecta have been reported till date; with no histopathological identification of the lesion.

A first case multiple idiopathic cystic lesions in jaw bones in a patient with osteogenesis imperfecta was reported by Jones AC in 1993. [8] A single mandibular cyst was reported in patient with osteogenesis imperfecta by Vorast Hin 2000. [9]

Dentigerous cyst is the most odontogenic developmental cyst of jaw bones, no association has been reported between the dentigerous cyst and osteogenesis imperfecta till date to the best of our knowledge, and here we report the first case of multiple dentigerous cysts in a patient with osteogenesis imperfecta.

CASE REPORT

A 16 year old male patient reported to the department of oral medicine and radiology; Nair hospital dental college with complain of difficulty in chewing due to multiple missing teeth (i.e. non eruption of permanent teeth following exfoliation of deciduous teeth). The patient gives the history of root canal treatment with mandibular left first molar around 3 years back following pain and swelling and silver amalgam restoration with maxillary right first molar and maxillary left second molar due to caries. No history of dental extraction was given by the patient. On checking past medical documents following past medical history was revealed,

1. Fracture of the left humerus at the time of delivery.
2. Tuberculosis at 3 months of age for which medication was taken for 6 months.
3. Fracture of the left humerus at the age of 2 years after fall from approximately 2 to 3 feet height
4. Fracture of the right humerus at the age of 5 years after fall from approximately 4 feet height.
5. Ophthalmology examination revealed blue sclera.

No significant family history of similar signs and symptoms was given by the patient.

Figure 1: Extra oral examination showing large head size, maxillary hypoplasia, mandibular Prognathism.

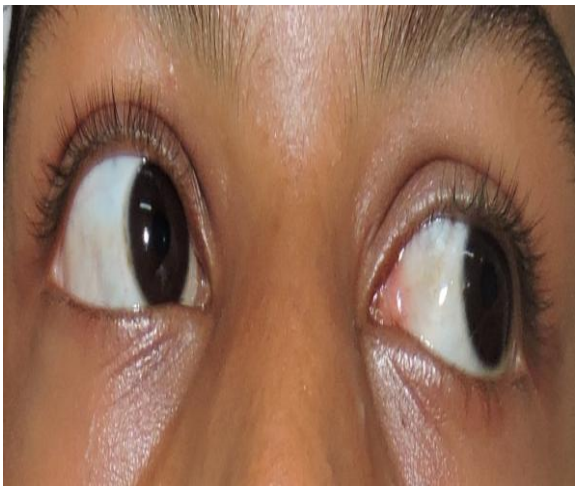


Figure 2: Blue sclera

On general examination, the patient was well oriented in space and time, lean in appearance, short stature, normal gait with satisfactory vital signs, height of 132 cm, weight 39 kilograms and normal intelligent

quotient. On extra oral examination (Figure 1) there was large head size, maxillary hypoplasia and mandibular prognathism. Blue sclera was confirmed on ophthalmological examination (Figure 2).

Intraoral examination (Figure 3A, 3B) showed

a. Missing maxillary right canine, maxillary left second and third molar, maxillary left central incisor, maxillary left canine, maxillary left second and third molar, mandibular left second premolar, second and third molar, mandibular right second premolar, first molar, second molar and third molar.
b. Over retained deciduous canines
c. Eruption bulge over the labial gingiva in the maxillary anterior edentulous ridge region
d. Silver amalgam restoration with maxillary right and left the first molar, mandibular left first molar and
e. Edge to edge anterior bite.
f. Teeth present were normal in shape and size.

A workingdiagnosis of multiple impacted teeth in a patient of delayed milestone was made.

Clinical differential diagnosis was

a. Metabolic bone diseases like osteogenesis imperfecta, rickets and vitamin D deficiency.
b. Hormonal disorders like hypothyroidism, hypoparathyroidism, and pseudohypoparathyroidism.
c. Syndromes associated with impacted teeth like Cleidocranial dysplasia, Gardner's syndrome, Gorlin sedano syndrome, Yunis varon syndrome.

Panoramic radiograph (Figure 4) showed multiple impacted teeth with incompletely formed roots and well defined circular radiolucency of size approximately 2.8 cm in diameter surrounded by thin corticated borders attached to the neck of each impacted teeth; maxillary right canine and second molar, maxillary left central incisor, lateral incisor, canine and second molar, mandibular left canine, second premolar and second molar, mandibular right canine, second premolar and second molar. Maxillary left third molar, mandibular left third molar, mandibular right first and the third molar was missing. Deciduous canines were over retained. Other skeletal radiographs; skull, humerus, fibula, tibia, chest showed no significant radiographic findings. Lateral cephalogram revealed prognathic mandible (Figure 5).

Provisional radiographic diagnosis of multiple developmental cysts associated with impacted teeth was made. Differential diagnosis of multiple dentigerous cysts of central variety was made.

On laboratory examination, the hormonal assay was found to be normal, serum calcium level (8.4 gm %), serum phosphate level (4.7gm %), alkaline phosphate level (224U/L) were within normal limits.

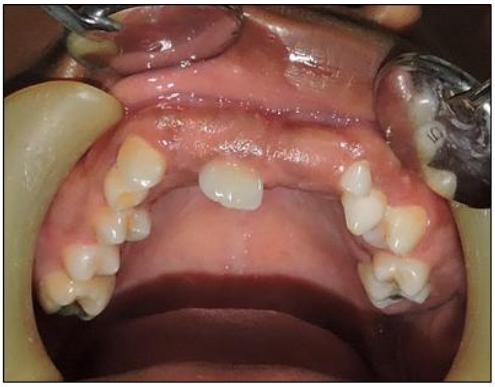
Based on history, clinical, radiography and laboratory findings a provisional diagnosis of multiple odontogenic developmental cysts associated with multiple impacted teeth in a patient of osteogenesis imperfect type 1 A was made.

Orthodontic consultation negated the opinion of orthodontic extrusion of impacted canines owing to poor bone density. Hence sectional surgical enucleation of cyst was planned so as to provide good healing period to the bone in view of metabolic bone disorder, cystic enucleation along with impacted teeth was done on the right side of mandible and tissue was sent for histopathological examination which showed fibrous connective tissue, composed of fibroblast and collagen fibres, odontogenic epithelium rests.

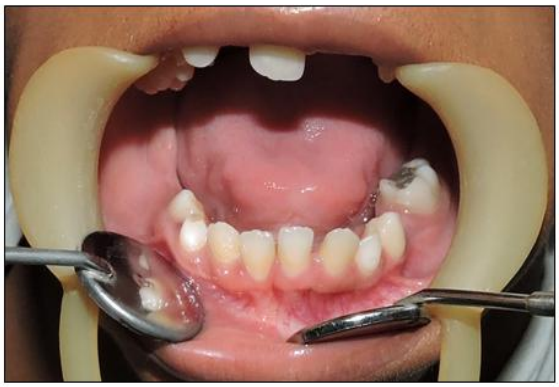
The final diagnosis of multiple dentigerous cysts in a patient of osteogenesis imperfecta types 1 A was made.

The patient was kept under follow-up for 1 year following a surgical procedure, panoramic radiograph showed incomplete

bone filling suggesting slow wound healing in our case even after long period of 1 year (Figure 6A, 6B).



3 A



3 B

Figure 3A: Maxillary arch showing missing maxillary right canine, second and third molar, maxillary left central incisor, and canine, second and third molar.
Figure 3B: Mandibular arch showing mandibular left second premolar; second and third molar, mandibular right second premolar, first molar, second molar and third molar and over retained deciduous canines.

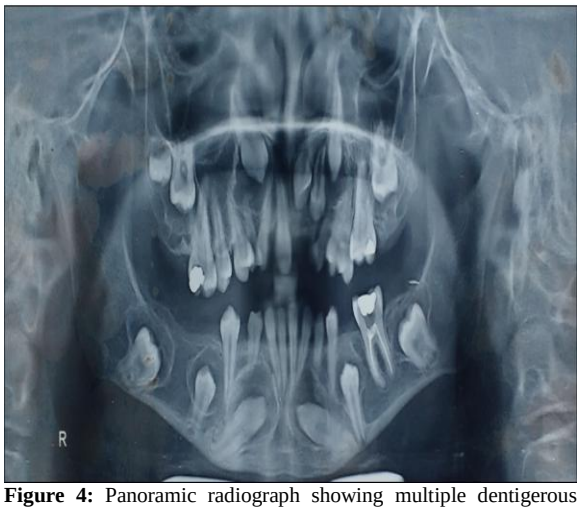


Figure 4: Panoramic radiograph showing multiple dentigerous cysts associated with impacted teeth.

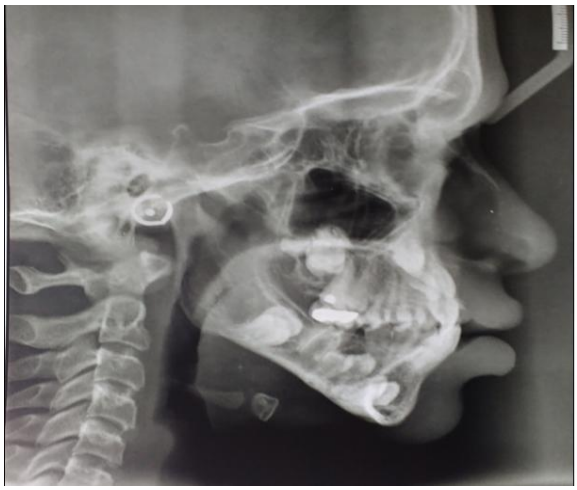
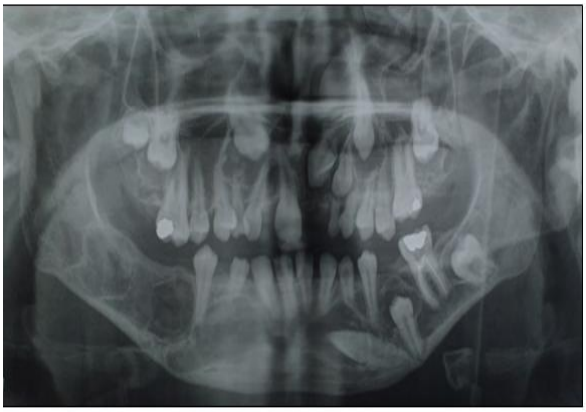
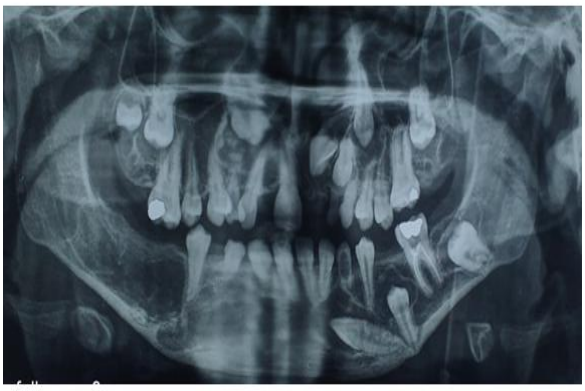


Figure 5: Lateral cephalogram showing prognathic mandible**.**



6 A



6 B

Figure 6A: Panoramic radiograph at follow up period of 3 months.
Figure 6B: Panoramic radiograph at follow up period of 1 year.

DISCUSSION

Osteogenesis imperfecta is the rare inherited disorder affecting the mesodermal tissue involving type 1 collagen, though non-inherited cases have also been identified. Osteogenesis imperfecta usually represent with brittle bones i.e. fractures with even slightest of trauma, in utero fractures in severe type, blue sclera, hearing loss, increased laxity of joints, poor muscle tone, kyphoscoliosis, in utero fractures, large head size, mandibular prognathism, maxillary hypoplasia, impacted molars, capillary bleeding and hearing loss.

The case reported here is the first case of multiple dentigerous cysts in a patient with osteogenesis imperfecta. Here diagnosis of multiple dentigerous cyst central variety in a patient of osteogenesis imperfecta type I A was made based on history and clinical, radiography and laboratory findings. Laboratory findings and hormonal levels within normal limits ruled out the association of any hormonal or metabolic bone disorder, which were being considered in differential diagnosis. Since no other feature of any other syndrome associated with multiple impacted teeth was seen in the patient, syndromic involvement was also ruled out.

In the case reported here, after surgical enucleation of cystic lesions in the right body of the mandible, reduced rate of healing of surgical wound with incomplete bone filling in some areas was noted during the follow-up period of 1 year. This finding was in contrary to the Vorast H, [9] who reported normal wound healing following surgical enucleation of single large mandibular cyst in apatient of osteogenesis imperfecta. The other case reporting multiple idiopathic mandibular cysts [8] in a patient with osteogenesis imperfect does not mention anything about healing aspect of the lesion post-surgical enucleation. Since not much data was available for comparison we hypothesized that incomplete bone healing in our case could be attributed to improper collagen synthesis and multiple and wide extents of the lesion as compared to the single cyst in previously reported the case.

Diagnosis of the Osteogensisimperfecta is usually done based on clinical and radiographic findings. [10] Collagen analysis of skin fibroblast can be done for confirmation.

In order to treat osteogenesis imperfecta multidisciplinary approach is required based on clinical manifestation, though no permanent cure has been identified till date, the aim should be to increase the density of bones so as to reduce the bone deformity.

CONCLUSION

Osteogenesis imperfecta is the rare genetic disorder with various craniofacial manifestations, possibly due to a mutation in multiple genes and thus requires a multidisciplinary approach for the management.

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