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Generalized idiopathic root resorption: a case report

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Background. Cases of generalized idiopathic root resorption have been reported in the literature. However the majority of previously reported cases have affected adults, and it has been rare that a possible link to a medical condition has been found.
Case Report. The subject of this case report was 11 years old when generalized root resorption was first diagnosed. This case report is unusual in that the resorption may be connected to a bone dysplasia. The possible medical diagnoses and links to tooth resorption have been discussed for this very rare case.
Conclusion. The investigation into the exact diagnosis of the patient and the treatment of tooth resorption is still ongoing and has required input from many disciplines of Medicine and Dentistry.

Introduction

Root resorption is broadly classified by site of origin as internal or external.

Both external and internal root resorption can be caused by trauma, orthodontic tooth movement, and may be a result of idiopathic resorption.

Internal root resorption may be caused by chronic inflammation of the pulp, restorative procedures, herpes zoster virus, cracked tooth syndrome, and invaginated cingulum[1].

The aetiology of external root resorption includes tooth reimplantation, impaction, periapical inflammation, pulpal necrosis, and pathological causes[2], such as ameloblastomas, giant cell tumours, fibro-osseous lesions, and oxalates in the blood[1].

External root resorption has also been reported to occur as a result of endocrine imbalances and systemic conditions, such as hypoparathyroidism, Paget's disease, Turner's syndrome, and hypophosphatasia[3].

Aetiological factors that may cause cervical external root resorption specifically include root planing, dento/alveolar and orthognathic surgery, and tooth bleaching[1].

Factors that have been implemented in the aetiology of both external and internal root resorption include trauma, orthodontic forces, and hereditary factors.

When there is no obvious cause for resorption, a diagnosis of idiopathic root resorption is made. This is often seen as starting at the cervical margin or apical region[1].

It is known that dentinoclasts are involved in the resorptive process. It has been suggested these cells may be activated by a number of agents, such as osteoclast-activating factor, macrophage chemotactic factor, prostaglandins, and bacterial products[4].

Histological examination of teeth affected by resorption show the presence of fibrous connective tissue filling resorption cavities and multiple osteoclasts in the resorption areas in pits or lacunae[5].

This case exhibits extensive root resorption of the permanent dentition in an 11-year-old girl. The possible rare diagnoses for this very unusual case are discussed.

Case report

A female aged 11 years and 7 months, was referred to the Eastman Dental Hospital in September 1999 by a hospital orthodontic department, regarding severe root resorption affecting several teeth.

The patient had a complaint of loose teeth, and there had been no previous history of trauma.

At the time of examination a diagnosis of rheumatoid juvenile arthritis had just been

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made. Previous medical history includes low birthweight and bilateral partial deafness since birth compensated by bilateral hearing aids.

There was no significant familial history.

The patient had been a regular attender to her general dental practitioner, and was recently referred to an orthodontist due to buccally displaced canines.

On extra oral examination the patient was tall, thin and had a slight lisp. There was no comment made on facial symmetry or proportion of facial features.

Intraoral examination revealed an early permanent dentition with all second molars present and partially erupted, grade 2 mobility of the lower right lateral incisor (42), and grade 1 mobility affecting the lower right central incisor and both the lower left central and lateral incisors (41, 31, 32). The upper central and lateral incisors (11, 12, 21, 22) sounded ankylosed on percussion.

The presence of an anterior open bite and buccally placed canines was noted. There were no carious lesions detected.

A panoramic radiograph and upper anterior periapical radiograph, which had been taken in May 1999 by the orthodontist, revealed resorption affecting most teeth (Figs 1 and 2).

Full mouth periapical radiographs were taken at time of exam (Figs 3 and 4) and confirm the presence of resorption affecting the following teeth:

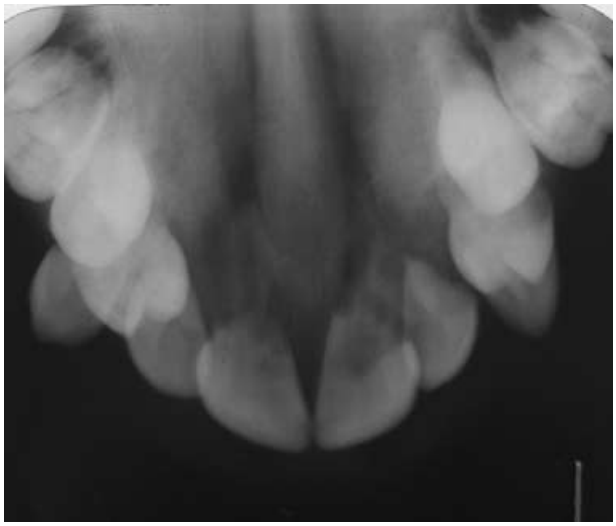

Fig. 2. Periapical radiograph showing resorption of the upper anterior teeth (May 1999).

6 3 2 1	1 2 3 4 6
6 5 4 3 2 1	1 2 34 5 6

(16, 13, 12, 11, 21, 22, 23, 24, 26, 31, 32, 33, 34, 35, 36, 41, 42, 43, 44, 43, 46).

As revealed by radiographic examination the resorption appeared to commence at the cementoenamel junction.

A decision was taken to review the patient again in December 1999. Main complaints at this review were increasing pain in the right knee and hands, deterioration of hearing on

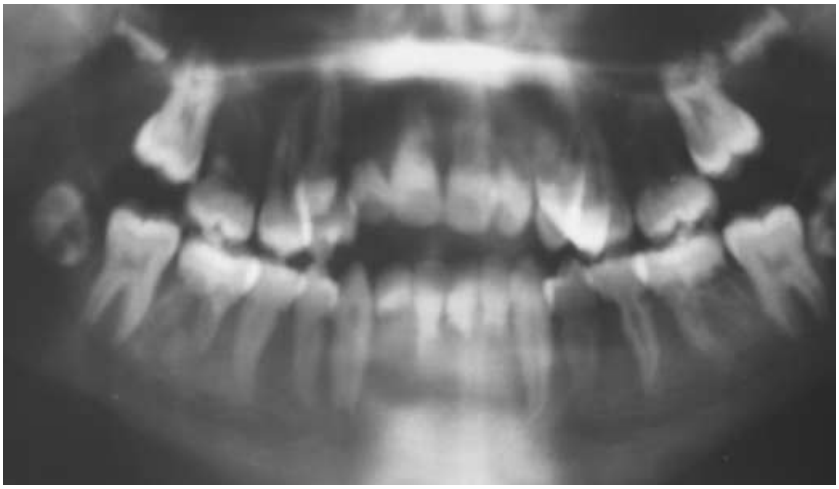

Fig. 1. Panoramic radiograph showing generalized root resorption of the permanent dentition (May 1999).

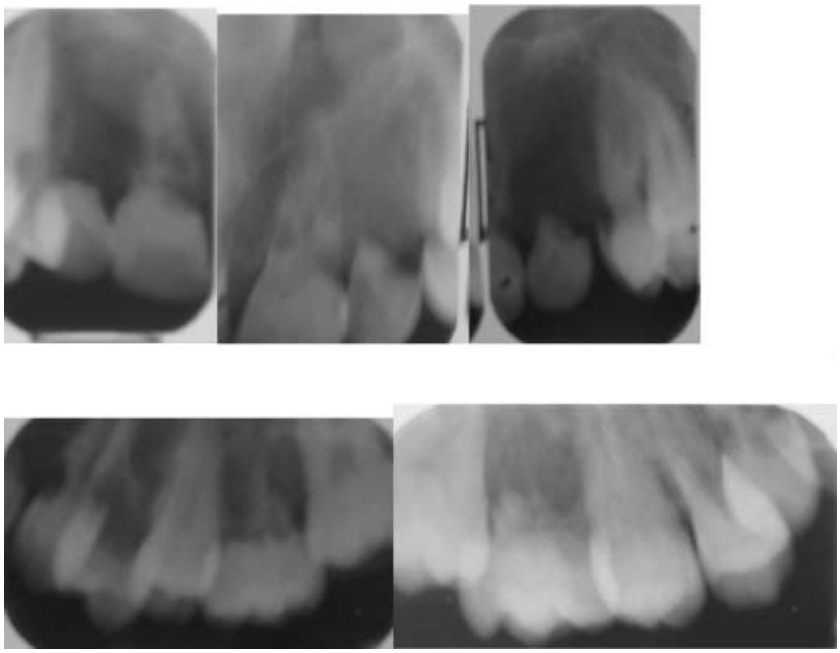

Fig. 3. Periapical radiographs showing continued resorption of the upper teeth (September 1999).

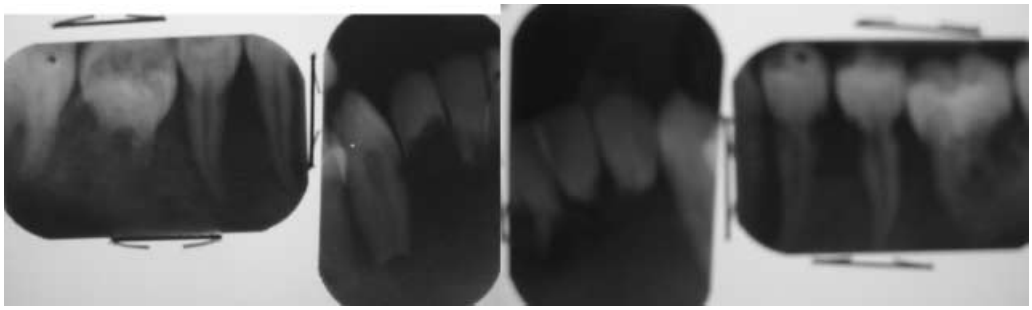

Fig. 4. Periapical radiographs showing continued resorption of the lower teeth (September 1999).

the right side, and the lower anterior teeth becoming increasingly mobile, especially the lower left lateral incisor (32).

A decision was taken to review again in 6 months' time as the patient was not keen on proceeding with extraction of loose teeth. In the meantime, it was decided to liaise with various consultants with regards to formulating a definitive diagnosis.

A consultant radiologist opinion was sought on all radiographs taken and a dental diagnosis of generalized idiopathic resorption was made. A thinness of all cortical structures of the surrounding bone was also noted, suggestive of hyperparathyroidism, although blood tests carried out by the medical team around the same time did not support this diagnosis.

The patient continued to visit the Eastman Dental Hospital for review appointments and in July 2002, main presenting complaints were severe tinnitus, complete deafness of the right ear, an increased stiffness affecting the shoulders, elbows and knees, and increasing difficulty when eating due to continued loosening of teeth.

Opinions were sought by a consultant implantologist, orthodontist, and restorative specialist with regards to the long-term management of the eventual loss of teeth. A treatment plan to provide immediate prosthesis following extraction was formulated, with a view to providing implants in the long term.

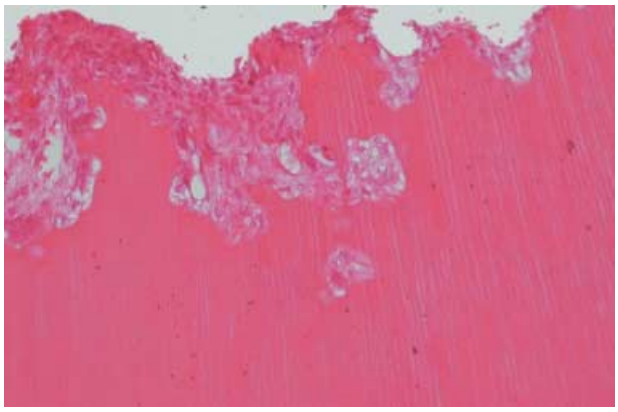

Fig. 5. Section of decalcified tooth showing large multinucleate osteoclasts in resorption pits.

The patient was still not keen on having any teeth extracted at this time but expressed great concern regarding her loose teeth. Mobility was now very marked, affecting upper and lower incisors as well as first permanent molars.

The lower right lateral incisor and lower right canine (42, 43) were splinted with glass ionomer in an attempt to stabilize them.

In January 2004, extraction of the following teeth were carried out under local anaesthesia and inhalation sedation (31, 32, 41, 42, 43, 44).

A lower immediate partial denture was fitted and lined with viscogel.

Histopathological examination of the extracted teeth showed a normal amelodentinal junction. Evidence of both internal and external resorption was found. The pulpal aspects of the dentine were lined by cellular fibrous tissue containing osteoclasts, many of which were found to be sitting in resorption pits. Trabeculae of bone, bacterial plaque, and a focus of inflammatory cells were also present (Fig. 5).

The histopathological features were consistent with idiopathic tooth resorption.

A panoramic radiograph taken in April 2005 revealed the continuing resorption of all teeth (Fig. 6). However, on clinical examination, some teeth were found to be only grade 1 mobile. Therefore, the decision was taken to continue to review rather than proceed to extraction of all teeth at once.

The prognosis of the remaining teeth is very poor. An upper mouth guard has been made to be worn at night to try and protect the remaining dentition, and the treatment plan is to extract as and when the teeth become very mobile with immediate replacement.

Around the same time when this patient was first seen by the dental team, she was also under the care of a consultant in metabolic medicine at a local hospital and had undergone various blood tests. The only abnormal result detected was a raised alkaline phosphatase

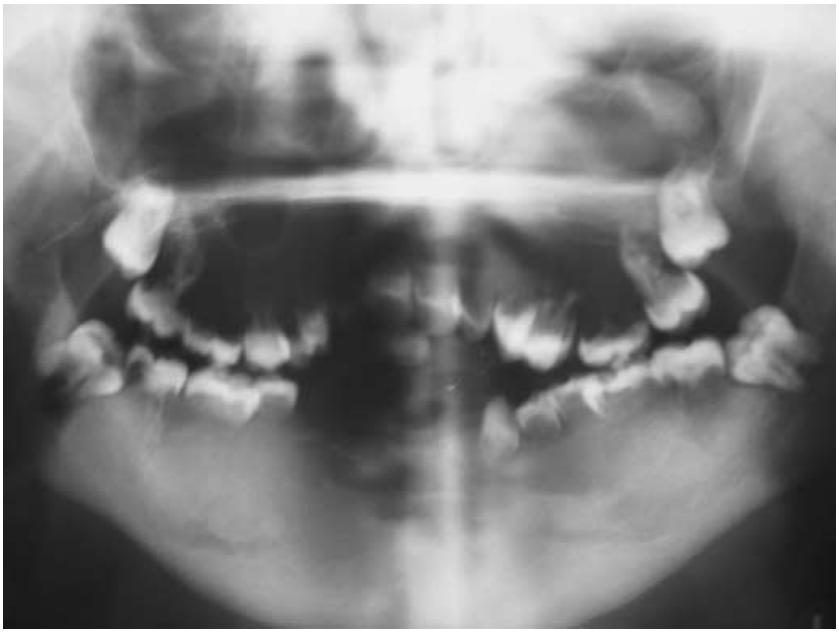

Fig. 6. Panoramic radiograph showing severe tooth resorption (April 2005).

at 508 U/L (normal range, 60–415 U/L). However, this result was not considered unusual for a child of this age as it was attributed to a growth spurt. Therefore, this result was not deemed significant enough for concerns to be raised by the medical team.

The diagnosis of juvenile arthritis that had been made very early on was now becoming unclear, due to other features that were inconsistent with this diagnosis, such as increasing deafness, bone fractures after minor trauma, absence of swollen joints, and normal blood results.

The patient was then referred to a specialist paediatric unit where she came into the care of a geneticist and specialist haematologist as well as having continued assessment by the metabolic medical team.

By March 2000, a likely diagnosis of polyostotic fibrous dysplasia was confirmed by the metabolic medical team. Fibrous dysplasia is a rare fibro-osseous lesion that can affect a single bone (monostotic fibrous dysplasia), or multiple bones (polyostotic fibrous dysplasia)[6]. Normal trabecular bone is replaced by a stroma of connective tissue with islands of calcified tissue.

It is now recognized that fibrous dysplasia is caused by a somatic activating mutation of the *GNAS1* gene that encodes for the GSα subunit of the G protein complex[7]. This results in an increase in cyclic AMP, which causes abnormalities in osteoblast differentiation. This in turn leads to the production of abnormal bone. There is also found to be an increase in interleukin 6-induced osteoclastic bone resorption[8].

Features of this disorder include increased risk of bone fracture and pain from symptomatic lesions. Fibrous dysplasia most commonly affects long bones, ribs, and the craniofacial bones.

Fibrous dysplasia of the maxilla or mandible may cause facial asymmetry, malocclusion, and loss of teeth as a result of fibrodysplastic bone[9].

Radiographically, jaw lesions are variable in appearance and there is a gradual transition from a normal uninvolved bone pattern to a cyst-like radiolucency and then on to a ground glass or orange peel appearance.

Displacement of teeth has been reported in fibrous dysplasia but root resorption is exceptional[10]. However, dysplastic changes in the dentine have been found with fibrous dysplasia. One such case was described by Wannfors *et al*.[6] in which a 26-year-old man with fibrous dysplasia was found to have irregularly shaped dentine tubules. It was suggested that the dysplastic changes in the dentine as well as bone may be due to a generalized defect in the mesenchymal hard tissue forming cells.

It is not known to the authors whether Paget's disease (juvenile type) was excluded at this time, as although Paget's disease is extremely rare below the age of 40, it does present with a raised blood alkaline phosphatase.

Other features of juvenile Paget's disease include increased bone formation and destruction, muscle weakness, bowing deformity of the lower limbs, non-traumatic fractures, and premature loss of teeth[11].

In 2002, a full skeletal survey and review by a specialist medical radiologist was carried out and diaphyseal dysplasia, also known as Camurati–Engelmann disease was suggested as a possible diagnosis.

Camurati–Engelmann disease is a rare type of 'symmetrical hereditary osteitis'. It is autosomal dominant and mostly begins in long bones causing thickening of the cortices, which eventually spreads to all bones.

Severe pain in the bones especially in the legs is a common feature as well as a waddling gait. Muscular hypoplasia is also a feature.

Caries has been associated with Camurati–Engelmann disease but there have been no reported cases of root resorption associated with this syndrome.

Other features of Camurati–Engelmann disease include increasing deafness (due to compression of the internal auditory canals as seen in this case), exophthalmos, optic nerve compression, diplopia, anaemia, and delayed puberty. The disease is usually self-limiting and the patient was not placed on any medication at this time. Medical care concentrated on possible surgical intervention if the bilateral deafness had worsened.

In July 2003, a high-resolution computed tomography scan of the base of the skull was

carried out at the local metabolic unit. The scan showed a ground glass appearance of the bone similar to that found in fibro-osseous dysplasia, but again the exact diagnosis still remained unclear.

The patient experienced five fractures during 2005 after minimal trauma, affecting the right and left humerus and left femur.

The authors referred the patient for genetic counselling and following this the patient was referred to an adult specialist metabolic unit for further investigation.

Since this report was first submitted in 2006, the genetic team to which the patient was referred to have suggested a new possible diagnosis of 'familial expansile osteolysis' (FEO) or a related condition of 'expansile skeletal hyperphosphatasia'.

Familial expansile osteolysis, also called 'hereditary expansile polyostotic osteolytic dysplasia', is a rare autosomal dominant bone dysplasia. It exhibits some histological similarity to Paget's disease in that the bone exhibits abnormal osteoblast and osteoclast function, but FEO has distinct enough features to be recognized as a unique condition[12].

The main clinical signs of FEO include early onset deafness (due to degeneration of middle ear ossicles), painful bony expansion with cortical bone thinning from focal osteolytic lesions within major long bones[13], and destruction of teeth.

The destruction of teeth seems to occur with extensive resorption starting at the cervical and apical region of the teeth[12].

Expansile skeletal hyperphosphatasia (ESH) is not a variant of FEO but is a related condition in that both diseases are allelic. ESH also exhibits early onset deafness, premature loss of teeth, and painful progressive hyperostotic widening of long bones, but there is absence of large osteolytic lesions.

The management of this case is still ongoing; S.H. may be considered for treatment with bisphosphonates and further DNA testing will hopefully establish the final diagnosis.

Discussion

Cases of multiple idiopathic root resorption have been reported in the literature. The majority of cases reported have involved adults. However, cases involving children have been reported. For example Snelgrove describes an incidental finding of generalized apical root resorption in a 14-year-old boy[2].

There has also been a case report involving two sisters aged 10 and 14 who both had multiple idiopathic root resorption of the permanent dentition[2].

In this case, resorption appeared to start at the cervical margins (Figs 1–4). This is typical of idiopathic root resorption along with resorption starting apically.

Also, in common with other reported cases of idiopathic tooth resorption[1,5,14], we found multinucleate osteoclasts in resorption areas on histological examination. It is interesting to note that this histological feature has also been demonstrated in bone from patients with monostotic and polyostotic fibrous dysplasia[15]. This has been a possible diagnosis for the patient in this case.

In addition to this, the CT scan carried out in July 2003 demonstrated a ground glass appearance of the skull bone, which is also seen in fibrous dysplasia.

Apart from fibrous dysplasia, diagnoses that have been suggested for our patient include juvenile arthritis and Camurati–Engelmann disease. Diagnosis of juvenile arthritis was rejected very early on.

A tentative diagnosis of Camurati–Engelmann disease was made on the basis of the patient's other signs and symptoms, such as increasing deafness and joint pain. However, S.H. showed no signs of delayed puberty or anaemia, which are other features of the disease.

There are no reported cases of any of the above conditions being linked to tooth resorption. Although the case study by Wannfors *et al.*[6] discussed above describes a finding of dysplastic changes in the dentine associated with fibrous dysplasia.

This case is atypical in several respects. Firstly, the presentation of features are not entirely typical of fibrous dysplasia or Camurati–Engelmann disease and diagnosis has been extremely difficult for all clinicians involved. Second, the nature of resorption affecting the dentition is extensive, and this may or may not be related to the patient's systemic condition.

Reinvestigation has now resulted in the possible diagnosis of FEO or ESH. Both have a typical presentation of destruction and early loss of teeth.

Olsen *et al*.[16], describes a similar case where both Mother and Daughter presented with progressive root resorption of the permanent dentition. This was linked to an unclassified bone dysplasia that showed features of FEO and hereditary hyperphosphatasia, and both patients had increased alkaline phosphatase levels.

In this case, however, there was no familial history and the raised alkaline phosphatase level found was attributed to a growth spurt.

It is not known to the authors whether later tests found the alkaline phosphatase levels to be abnormally raised.

The genetic team will be carrying out further testing to ascertain the exact diagnosis of this patient's condition. In addition to this she may be placed on bisphosphonate therapy.

With regards to S.H.'s dental management, the plan is to conserve the remaining dentition as long as possible with planned extractions of mobile teeth in the long term and provision of prosthesis.

If implants are to be considered the alveolar bone will need assessment. Implants may be contra-indicated if the patient is treated with bisphosphonates[17], as there is recent evidence to show that bisphosphonate therapy may play a role in the development of osteonecrosis of the jaws, although it is not certain whether this reason would be a contraindication in a young patient.

In addition to this the disturbed bone remodelling may also contraindicate implant placement. Therefore, before deciding whether implants could be a treatment option, a 'test fixture' would have to be placed.

Management of this case has been difficult and has required a multidisciplinary approach.

After years of investigation the definitive diagnosis of the patient's medical condition and, hence, the possible cause of extensive tooth resorption is still very much uncertain, and investigations are still ongoing.

The authors welcome any correspondence regarding similar cases.

What this paper adds?

- Provides knowledge on rare bone diseases and associated dental signs.
- Explores possible links between tooth resorption and metabolic bone diseases.
- Discusses the multidisciplinary approach necessary in the diagnosis and treatment of this rare case.

Why this paper is important to paediatric dentists?

- Discusses the process of investigation, and the possible causes of tooth resorption affecting the paediatric patient, which may aid in the diagnosis and treatment of similar cases.
- Highlights the different disciplines that may be needed in order to diagnose and treat rare cases such as this one.

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