VARIA

Generalised prepuberal periodontitis: literature review and case presentation

Samoilă Anca Irina¹, Răducanu Anca Maria², Cristea Irina³

București, România

Abstract

Generalized prepuberal periodontitis (GPP) is a rare and aggressive periodontal disease which has early onset and extremely rapid evolution. Thus the affected teeth, both primary and permanent, are early lost or extracted.

This disease is frequently associated with systemic diseases or a genetic predisposition to this illness, but the determining factor is the periodontal pathogenic anaerobic microflora.

It is presented a case of a little boy (15 month old) with PPG. Based on the oral features, typical for PPG, an extremely severe systemic disease (X histiocytosis the disseminated acute type or Letterer-Siwe disease) was diagnosed.

Introduction

GPP is a rare, aggressive periodontal disease with the first signs appearing early in the childhood, characterized by a rapid evolution. [1, 4] Due to these facts it is considered to be the most severe form of prepuberal periodontitis. [1, 4]

It affects both deciduous and permanent teeth of young children. The evolution of this disease leads to the early loss of these children's teeth. [1]

Etiology

The etiology of this disease is more likely to be associated with immune system deficiencies and chromosomal disorders, rather than with the presence of dental plaque or calculus. Anaerobic microorganisms, such as: Actinobacilus Actinomycetem Comitans, Bacteroides gingivalis, Bacteroides intermedia, are being frequently revealed in the periodontal pockets of the affected patients. [1, 2, 5, 6] Functional deficiencies of immune cells such as diminishing of phagocytosis, and of the leucocytes adhesions, are involved in the disease evolution. [2, 7]

It has been recently discovered that PP has appeared under the influence of a gene situated on chromosome 11q14. Chromosome 11q14 contains the gene of cathepsin C, which is responsible for the appearance of certain serious general illnesses such as: Papillon-Lefevre Syndrome and Haim-Munk Syndrome, diseases accompanied by severe periodontal suffering. [1, 5]

GPP can also appear as a sign of general illnesses such as: leukaemia, neutropenia and X histiocytosis, sometimes helping to diagnose these systemic illnesses like in our clinical presented case. [5, 6]

X histiocytosis is characterized by the neoplasic growth of the Langerhans cells and of the eosinophiles. [2, 7]

The causative factors of this illness are unknown, but immunological and genetic disorders, viruses and bacteria are supposed to play a part in its etiology. [2,3,7]

¹ Lecturer, Department of Paediatric Dentistry, Faculty of Dental Medicine, University of Medicine and Pharmacy "Carol Davila" Bucharest, Romania

² Associate Professor, Department of Paediatric Dentistry, Faculty of Dental Medicine, University of Medicine and Pharmacy "Carol Davila" Bucharest, Romania

³ Assistant Professor, Department of Paediatric Dentistry, Faculty of Dental Medicine, University of Medicine and Pharmacy "Carol Davila" Bucharest, Romania

The acute disseminated form (Letterer- Siwe disease) affects small children and is clinically manifested through hepatosplenomegaly, diffuse bone lesions of the skull, jaws, long bones, skin lesions, enlarged lymph nodes, otitis, mastoiditis and respiratory tract infections. Bone lesions of the jaws produced by the destruction of the alveolar bone have as a consequence the exfoliation of the teeth or of the dental germs. [3] This X-ray aspect is also known as "floating teeth". [7] Gum swallowing and necrosis also appear. The prognosis of the disease is reserved it is often a lethal illness (60%). [7,8]

Clinical features

GPP is the most severe clinical form of prepuberal periodontitis this being determined by the number of affected teeth and by the gravity of the clinical manifestations.

The disease has an early start (2-4 years), soon after the eruption of the primary teeth and its progression is extremely rapid. [1]

The clinical picture of GPP is made out of progressive destruction of the bone and forming of deep pockets. [7] The teeth have various degrees of mobility and migration; they are soon to be lost either by extraction or expulsion. [6, 7] GPP is accompanied by moderate plaque, calculus deposits and gingival swallowing. [5, 7] The functional disorders (chewing, physiognomy, phonetics) complete the clinical picture. [1]

The X-ray shows severe bone destruction and the presence of deep pockets with the appearance of the "floating teeth" aspect. [7]

Treatment

The success of the treatment of the PP depends on early on the diagnosis and is directed to the therapeutic attack on the infectante microorganism and providing a healthful, free atmosphere of infections. [1] The treatment depends on early diagnosis and on the severeness of the illness at this moment and consists of:

- 1. Local treatment
 - Decrease the level of microflora and of bacterial plaque [1, 5] by:
 - prophylaxis every 3 months (correction and control of oral hygiene and professional brushing) [1, 5]

- local subgingival irrigations with clorhexidine, hydrogen peroxide or/and tetracycline elixir 250 cc [1,5]
- Tooth scaling and root planning [1]
- The extraction of the irreparable teeth [1]
- Recall every 3 month, to avoid bacterial insemination of other teeth [1]

2. General antibiotherapy (penicillin, erythromycin or doxycylin) by 5 days

- Tetracycline 250 mg., after 8 years old, 25-50mg./kg body weight/day, for 7 days. The permanent teeth can display staining and also have the increases risk of presenting oral candidiasis. [1,5]
- Amoxicillin 20-40 mg./kg body weight/day, and Metronidazol 15 mg./kg body weight/day for 7 days or Augmentin suspension 25 mg./kg body weight/day. [1,5]

Case presentation

It is presented a little boy, MN, 15 month old, with GPP. Based on the GPP typical oral features, it has been diagnosed an extremely severe systemic disease (X histiocytosis), the acute disseminated form (Letterer- Siwe disease).

The first time the patient came at the Department of Paedodontics, he was complaining of gingival pains and bleeding and changes in the teeth position.

The mother said that the patient had had repeated upper respiratory infections and several accidental falls. The general practitioner and the dentist from his home village believed that this falls determined unhealed dental injuries, still responsible for the symptoms.

The repeated blood tests indicated normal results. The examination revealed the following oral and general manifestations:

- 1. General manifestations:
 - ill-being with low fever, fatigue and anorexia.
 - latero-cervical and submandibular adenophaty
 - muscular weakness that had provoked the frequent accidental falls.
- 2. Oral manifestations:
- massive alveolar-bone destructions with deep pockets (Fig. 1, 2)



Fig. 1, 2. Gingival inflamation, deep pockets, teeth migration, dental plaque deposits



Fig. 3 Expulsion of 1.1 situated oral from the temporary incisors

- dental mobility (2-3 degree)
- teeth migrations (Fig. 1, 2)
- acute proliferated gingival inflammation with congestion and areas of necrosis
- spontaneous and provoked gingival bleeding
- spontaneous gingival pains, as well as provoked
- deposits of dental plaque and calculus (Fig. 1, 2)
- the exfoliation of 75 and 85, although they looked like erupting teeth
- the expulsion of 11, situated oral from the temporary incisors. The tooth looked like a bone arrestment but it had the shape and size of a normal permanent incisor (Fig. 3)
- the Rx confirmed the severe alveolar bone destruction and the typical PPG aspect of floating teeth (Fig. 4)

After anamnesis, oral, facial and general examination a severe general illness was suspected. This could have been the cause of the periodontal disease.

After the removal of the dental plaque and calculus the mother was instructed how to provide a better oral hygiene. Because of the severe oral lesions the patient was referred to the Children Department of the Fundeni Hospital from Bucharest, there it was determined that the patient was suffering of: Letterer-Siwe diseases, final state. After 1 month the patient passed away.

Discussions and conclusions

PP It is an extremely destructive rare disease for the periodontium and of fast evolution. It reaches the deciduous teething, usually soon after the eruption of the teeth. Correlation almost always exists with systemic problems and genet-



Fig. 4. Rx - severe alveolar bone destruction

ic inheritance of predisposing characters, but the contact with the periodontal pathogenic microflora is the primary factor in the process of destruction of the disease. The precocious diagnosis and the basic periodontal treatment, associated to the antibiotic therapy, are the main weapons for the combat this disease.

References

1. Contreras Genny Durán, Periodontitis prepuberal criterio, diagnostico, patogenia y terapeutica, *Revista Odous Cientifica*, 2000, **1**(1), http://servicio.cid.uc.edu.ve/ odontologia/revista/ v1n1/1-1-3.pdf

2. Einwag J., Pieper K., Parodontopathien, in *Kinderzahnheilkunde*, Urban & Schwarzenberg, 2002, **16**:335-336.

3. Gisela Estela Rapp, Ana Carolina Fragoso Motta, Periodontal Disease Associated with Langerhans' Cell Histiocytosis: Case Report, *Brazilian*

Correspondance to: Samoilã Anca Irina, Str. Ionel Perlea 12, tel. 314.20.80/132 E-mail samoilaanca@yahoo.com

Dental Journal, 2000, 11 (1):59-66.

4. Hart TC, Hart PS, Bowden DW, Michalec MD, Callison SA, Walker SJ, Zhang Y, and Firatli E. Mutations of the cathepsin C gene are responsible for Papillon-Lefèvre syndrome, *Journal of Medical Genetics*, 1999; **36**: 881-887.

5. McDonald R.E., Avery D.R., *Dentistry for the child and adolescent: Gingivitis and Periodontal Disease*, Mosby, Seventh Edition, 2004, **20**:458.

6. Martinez B.A., Ruiz F.E., Periodontal diseases as bacterial infection, *Avances en Periodoncia e Implantología Oral*, 2005; **17**(3):111-118.

7. Pinkham J. R., Casamassimo P.S., H.W. Fields, McTigue D. J., Nowak A.J: *Pediatric Dentistry: Periodontal Problems in children and Adolescents*, Elsevier Saunders, Fourth Edition, 2005, **24**:421.

8. Sedano O. Heddie, Premature loss of teeth, Periodontics Information Center, UCLA School of Dentistry, http://www.dentistry.ucla.edu/ PIC/visitors/Teethloss/page1.html