Aggressive periodontitis of the primary dentition associated with idiopathic immune deficiency: case report and treatment considerations

Enrique Bimstein* / Michael McIlwain** / Joseph Katz*** / Greg Jerrell**** / Robert Primosch*****

The present case, of a child with an idiopathic immune deficiency and aggressive periodontitis in the primary dentition, serves as an example for the treatment considerations in these cases. Extraction of all the primary teeth proved to be the most adequate treatment. It allowed the child to eat properly and prevented unwanted infections that could endanger the life of the child. The newly erupted permanent teeth have been subjected to careful oral hygiene, clorhexidine topical applications, and have mild gingival inflammation and no attachment loss.
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INTRODUCTION

Immune deficiencies are a heterogeneous group of rare disorders characterized by the decreased ability of the immune system to fight infections, abnormal leukocyte function, inability to get neutrophils and monocytes to leave the blood vessels to the sites of injury and bacterial challenge, and decreased cellular adhesion.[1-3] Aggressive gingival and periodontal diseases, with rapid and extensive alveolar bone loss that leads to early exfoliation of primary teeth, are significant and dramatic findings in children with immune deficiencies.[4-6] The clinical and radiographic appearances of these diseases can resemble generalized aggressive periodontitis[7] in the primary or permanent dentition, that were classified in the past as generalized prepubertal or generalized juvenile periodontitis.[8,9] Other reports of oral findings in these patients include stomatitis, ulcerations of the mucous membranes, and facial cellulitis.[10-11]

Cases of children with immune deficiencies, including those with leukocyte adhesion deficiency (LAD), involving periodontal disease have been reported in the literature.[2,4,11] However, further information on the treatment considerations of these cases is required. The purpose of the present manuscript is to discuss the local and systemic considerations of the treatment of severe periodontitis in children with immune deficiencies, based on a two year follow-up of a child, who had idiopathic immune deficiency.

CASE REPORT

A 4-year-old Caucasian male child (JT) was first seen by the authors at the Pediatric Dentistry Department of the University of Florida College of Dentistry for a re-evaluation of gingival and periodontal diseases. The mother reported that LAD was recently suspected, that 3 primary anterior teeth exfoliated "with long roots", and that the child complained of severe pain, while eating due to the increased tooth mobility. JT's past medical and dental history indicated that he was diagnosed at age 3-years with Idiopathic Immune Deficiency consisting of a granulocyte defect characterized by defective bacterial killing, reduced chemotaxis and neutropenia. At that time, JT:

* Enrique Bimstein, CD, (Cirujano Dentista, México), Professor, Department of Pediatric Dentistry, University of Florida College of Dentistry, Gainesville, Florida and in the Hebrew University in Jerusalem.
** Michael McIlwain, DMD, Resident, Department of Pediatric Dentistry, University of Florida College of Dentistry, Gainesville, Florida.
*** Joseph Katz, DMD, Professor, Department of Oral and Maxillofacial Surgery and Diagnostic Sciences, University of Florida College of Dentistry, Gainesville, Florida.
**** Greg Jerrell, DDS, Associate Professor, Department of Pediatric Dentistry, University of Florida College of Dentistry, Gainesville, Florida.
***** Robert. Primosch, DDS, MS, MEd, Professor, Department of Pediatric Dentistry, Associate Dean for Education, University of Florida College of Dentistry, Gainesville, Florida.

Send all correspondence to Dr. Enrique Bimstein, Department of Pediatric Dentistry, University of Florida College of Dentistry, P. O. Box 100426, Gainesville, FL 32610-0426.

Voice: 352 392 4131
Fax: 352 392 8195
E-mail: ebimstein@dental.ufl.edu

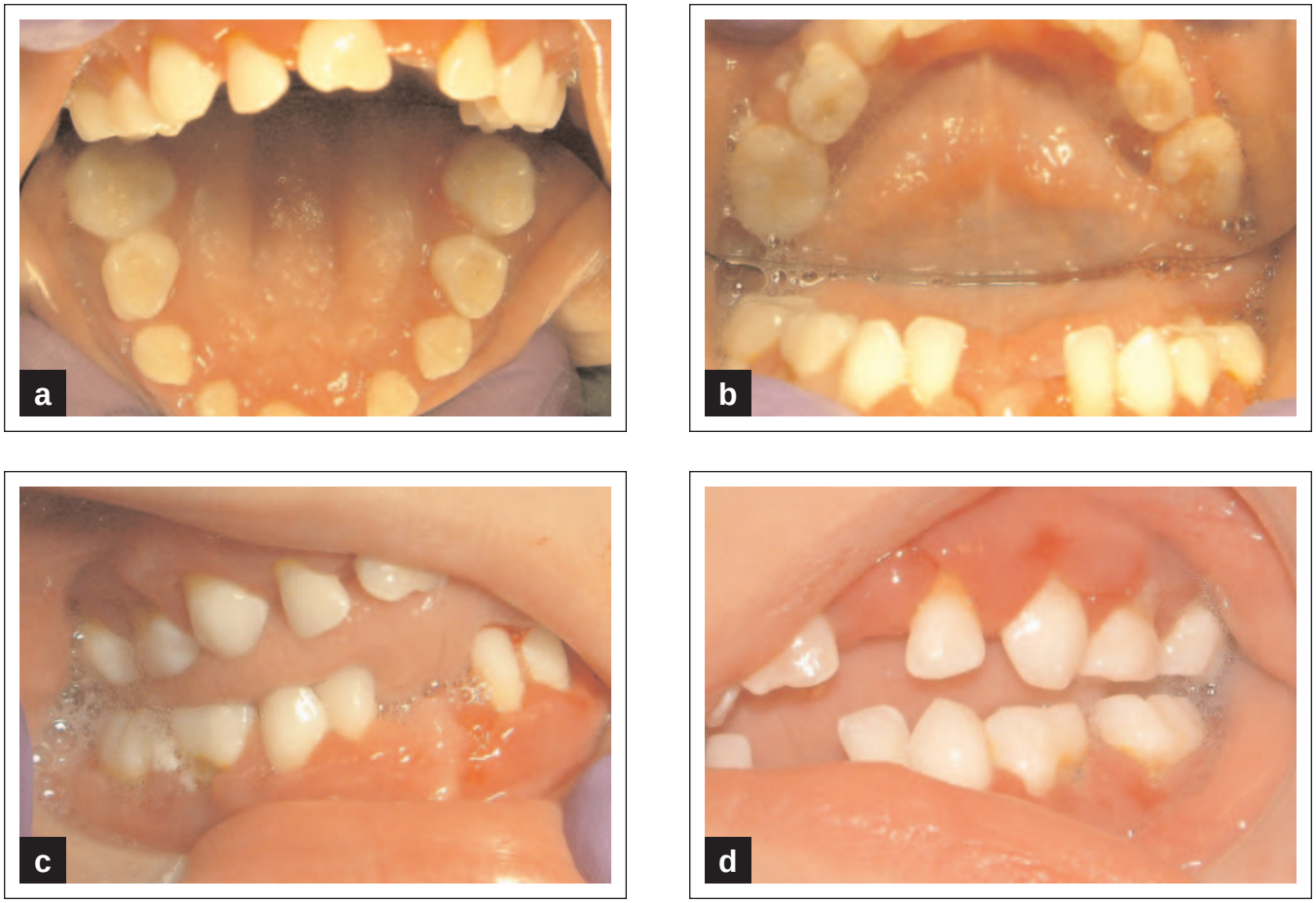

Figure 1. Clinical pictures of a child with aggressive periodontitis in the primary dentition related to immune deficiency. Severe gingival inflammation and recession are evident.

a. Maxillary occlusal view (mirror view).
b. Mandibular occlusal view (mirror view).
c. Right side.
d. Left side.

a. had a developmental delay (18-24 months development), normal weight (16.5 kg), short stature of 37 inches (below the 5 percentile), and delayed speech development.
b. was under chronic antibiotic therapy for recurrent systemic infections facilitated by the immune deficiency.
c. hematological workup included slight leukopenia of 4.5 thou/cu mm (normal range 5.5-15.5 thou/cu mm) with severe neutropenia of 4.9 % (normal range 30-85%), normal RBC with anisocytosis, microcytosis with hypochromia. The sedimentation rate was elevated after the first hour to 61 mm/h.
d. oral clinical examination revealed severe gingivitis with bleeding easily provoked by touch and spontaneous bleeding. Candidiasis, which increased the severity of the gingival inflammation, as the result of the chronic antibiotic treatment was suspected. Therefore, ketoconazole 100 mg (0.5 mg/5 c. c.) four times a day for 14 days was prescribed. Twelve days later, the mother reported that ketoconazole was not effective in reducing the gingival inflammation. In an additional attempt to reduce the gingival inflammation, dexamethasone (Decadron) gel was prescribed six times a day for 14 days. However, this treatment also proved to be ineffective in reducing the gingival inflammation.

Five months after the initial examination, an oral assessment revealed erythematous and hyperplastic gingiva. Therefore, the following recommendations were provided: a) improve oral hygiene; b) chlorhexidine rinses (Peridex); c) recall examination every 3 months. A week later, the mother indicated that the child had been diagnosed with agranulocytosis and was prescribed a chronic coverage with a broad spectrum antibiotic (Cipro 250 mg/day) and received granulocyte colony stimulating factor (10 mg./kg/day).

At age 4-years, when first seen by the authors, JT's clinical examination revealed missing teeth (F, O and P), severe gingival inflammation and recession in the remaining primary teeth (Figures 1a-d), and increased abnormal mobility that elicited pain during

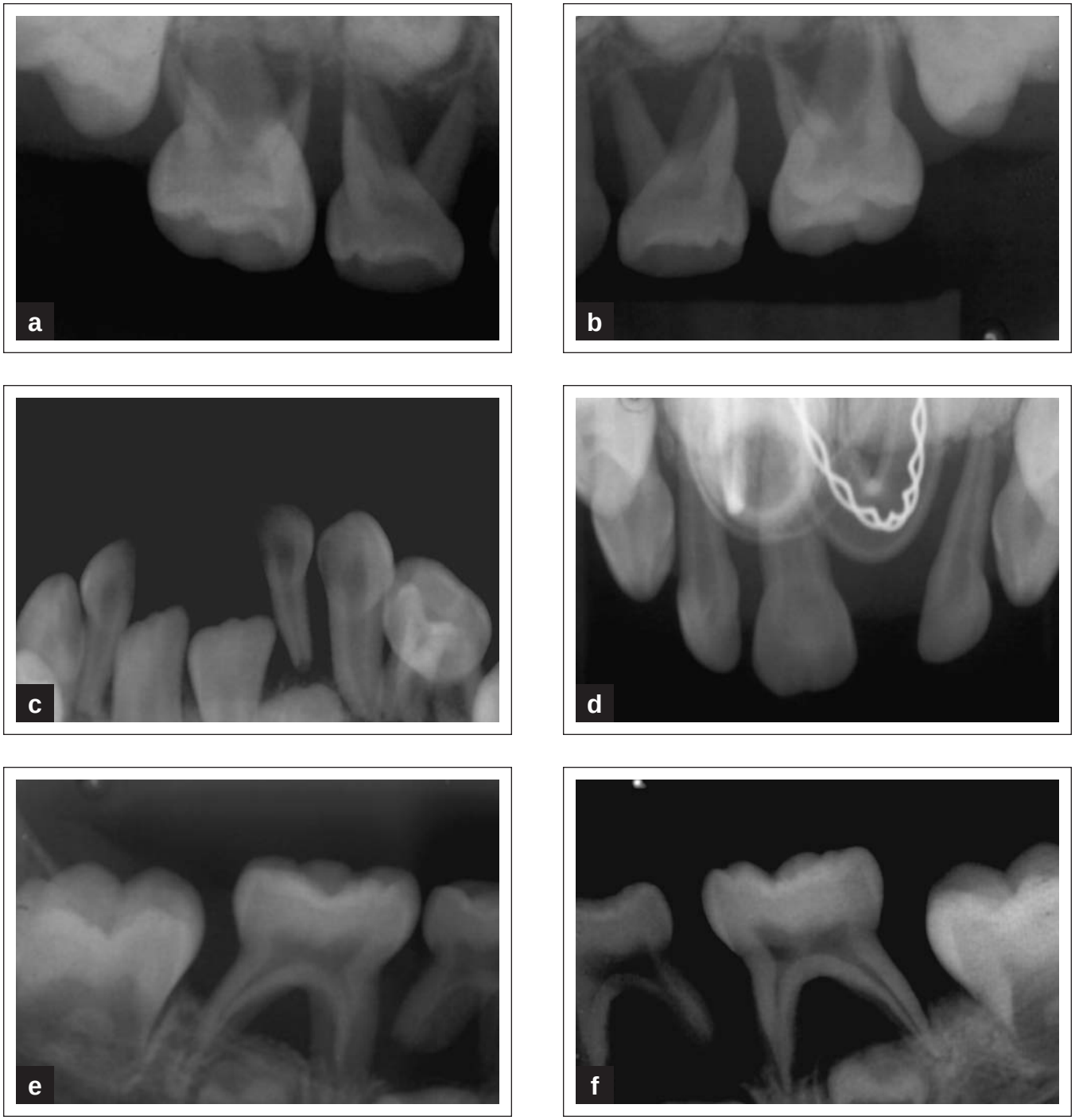

Figure 2. Radiographs of a child with aggressive periodontitis in the primary dentition related to immune deficiency. Severe alveolar bone loss is evident in all quadrants.
a. Maxillary right molars.
b. Maxillary left molars.
c. Mandibular anterior area.
d. Maxillary anterior area.
e. Mandibular right molars.
f. Mandibular left molars.

the examination. During the examination and a toothbrush prophylaxis, JT showed strong opposition. It was decided to perform a radiographic examination and extract his primary teeth under general anesthesia.

The immunologist was contacted to discuss the medical history, precautions that needed to be taken for surgery, and the type of antibiotic therapy required to prevent the development of local or systemic infec-

tions after the extractions. A post operative course of Clindamycin for two weeks, 30mg/kg/day divided in three doses, was recommended. In addition, it became evident that JT had neither LAD I, since it has been discovered that he expresses CD18 integrin, nor LAD II since he had no Bombay phenotype.[3,5,12]

A month later, following informed consent, a course of granulocyte activating factor and antibiotic prophylactic therapy, the dental radiographic examination and treatment were done under general anesthesia; the radiographs revealed extensive alveolar bone loss in all four quadrants (Figures 2a-f). Therefore, all the remaining 17 primary teeth were extracted and the recommended post operative antibiotic therapy was prescribed.

A two-month post operative evaluation revealed healthy oral soft tissues, along with the beginning of the early eruption of the maxillary first permanent molars (Teeth 3 and 14) and mandibular permanent central incisors (Teeth 24 and 25). JT's mother indicated that extraction of the primary teeth resulted in a major improvement in the nutrition, since he was relieved of the pain elicited from the inflamed tissues and increased tooth mobility. She also indicated that JT developed his "own methods" to cut and chew food it in a satisfactory manner. The mother was instructed to perform cotton swab clorhexidine cleaning around the erupting permanent teeth twice a day.

Nine months after the extractions, while the child was hospitalized for a routine exam, the mother reported that JT continued with no oral complaints and that she has been doing the clorhexidine topical cleaning. An oral clinical examination revealed the erupted permanent teeth with minimal gingival inflammation, normal mobility and no attachment loss.

DISCUSSION

Cases of aggressive periodontal diseases affecting primary teeth have been described in children with or without systemic diseases or syndromes such as LAD, neutropenias, Papillon Lefèvre Syndrome, Chédiak-Higashi Syndrome, hypophosphatasia, etc.[13-17] However, the appearance of severe periodontitis affecting the primary teeth has been mostly related to systemic diseases.[18] Therefore, when severe periodontitis is found in primary teeth, the clinician should refer the patient for a thorough systemic evaluation.

Clinicians, who confront children with aggressive periodontitis in the primary teeth, must decide between a conservative treatment approach avoiding extractions, or a radical treatment approach that involves extractions. In general, aggressive periodontitis in primary teeth may be successfully treated by a conservative approach that includes local (scaling, curettage, antiseptic rinses and root planning) and systemic therapy (antibiotic) in patients with good compliance.[16,17] However, each case must be considered individually based on the severity and extent of the gingival and periodontal diseases, the systemic condition of the patient, behavior management considerations and compliance.

In the present case, analysis of these variables contraindicated a conservative approach since:

1. **The clinical and radiographic examination revealed early exfoliation** of primary incisors, generalized severe gingival inflammation and recession (Figures 1 a-d), generalized aggressive alveolar bone loss (Figures 2a-f) that included the resorption of the lamina dura surrounding several permanent tooth follicles (Figures 2a, b, c, e, f) endangering the developing tooth germs,[19] and severe pain easily elicited by tooth mobility.
2. **An idiopathic immune deficiency was present.** This was a most significant variable in this case, since the seriousness of the disease was evidenced by the ineffectiveness of repeated antibiotic therapy for systemic infections, and of ketoconazole and dexamethasone in the treatment for the oral infections. Furthermore, in cases with immune deficiencies, there is a large mortality prevalence related to overwhelming sepsis, despite antibiotic therapy.[1-4,11,20,21] Therefore, a persistent effort to maintain the infected primary teeth in the present case could have endangered the patient's life, as it has been previously indicated in a case of LAD.[2] In addition, the possibility of bacteremia subsequent to extraction in the present case required antibiotic coverage, and the extraction of all the primary teeth at one appointment permitted antibiotic coverage to occur only once.
3. **Behavior management considerations.** The ability of the child to cooperate, and allow for repeated treatments in a clinical setting, is required for a conservative approach. On the other hand, when the child is not able to cooperate even with sedation as in the present case, treatment under general anesthesia and a radical approach involving a one time treatment is indicated.
4. **Compliance**. Compliance is essential in the treatment and prevention of recurrence of infectious diseases. In the present case, compliance was not a factor to be taken into consideration during the initial treatment, due to the presence of severe gingival and periodontal diseases and the systemic condition. On the other hand, at present and in the future, considerable effort will be required to prevent the establishment of periodontal diseases in permanent teeth. A prevention regimen included regular oral hygiene, frequent professional dental prophylaxis and chlorhexidine rinses. The outcome of this strategy proved to be successful in a case of Behcet disease,[6] which is also an autoimmune disease, and appeared to be successful in the present case, seven months after the eruption of the permanent teeth. However the prognosis for a long period of time is uncertain.

The present case emphasizes the need for cooperation between medical and dental professionals, and the substantial significance of the prevention or early treatment of gingival and periodontal diseases that are related to systemic conditions. Once these diseases are established, the well being of the patient is the crucial parameter in the treatment decision process.

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