Chronic idiopathic neutropenia associated with gingival enlargement

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A girl with chronic idiopathic neutropenia who developed gingival enlargement at seven years of age is presented. Intraoral examination revealed generalized gingival inflammation with a tendency to bleeding and inflammatory gingival enlargement localized to the anterior region. A considerable amount of bacterial plaque was noted on the teeth. There were also 4-5 mm pocket depths around the first molars. Radiographic examination also indicated the presence of incipient bone loss around the first molars in both jaws. The patient, who was diagnosed as localized prepubertal periodontitis with generalized gingival inflammation and anterior gingival enlargement, accentuates the importance of evaluation of periodontal status in patients with chronic idiopathic neutropenia, to avoid the destruction of supporting structures of the dentition.

Key words: neutropenia, gingival enlargement, prepubertal periodontitis.

The most frequent hematological disease associated with gingival enlargement is accepted as leukemia. However, in patients with immunodeficiency, as in neutropenia, neutrophil dysfunction and aplastic anemia[1-8], periodontal manifestations ranging from-marginal gingivitis to rapidly progressive periodontitis may be prominent[5-8]. Sometimes stomatitis with ulceration is the only major clinical manifestation of neutropenia[7]. This presented case describes localized prepubertal periodontitis with generalized gingival inflammation and anterior gingival enlargement in a seven-year-old girl with chronic idiopathic neutropenia.

Case Report

A 4 ½ -year-old girl was referred to the Hematology Unit because of recurrent upper respiratory tract infections, tonsillitis, pneumonia, sinusitis, and sinopulmonary infections since one year of age, and was treated for pulmonary tuberculosis. Her white blood cell count (WBC) ranged between 3.5-6.5 x 10^9/L and absolute neutrophil count between 0.18-0.58 x 10^9/L. The bone marrow aspiration revealed arrest in promyelocyte maturation. The immunoglobulin levels and lymphocyte subsets were normal. There was fourth degree consanguinity between the parents; however, neither the parents nor her two siblings had neutropenia. Her mother complained of recurrent aphthous lesions. She was treated with granulocyte colony stimulating factor (G-CSF) for 10-20 days during infections, which recurred every 15-45 days with the diagnosis of chronic idiopathic neutropenia. When she was seven years of age, gingival enlargement was observed and she was referred to the Periodontology Department of the Faculty of Dentistry for further evaluation.

Intraoral examination revealed abnormal teeth relations and generalized gingival inflammation with a tendency to bleeding and inflammatory gingival enlargement localized to the anterior region (Figs. 1, 2). A considerable amount of bacterial plaque was noted on the teeth. There were also 4-5 mm pocket depths around the first molars. Accordingly, radiographic examination indicated the presence of incipient bone loss around the first molars in both jaws (Fig. 3). On the basis of clinical and radiographic findings, the patient was diagnosed as localized prepubertal periodontitis with generalized gingival inflammation and anterior gingival enlargement.

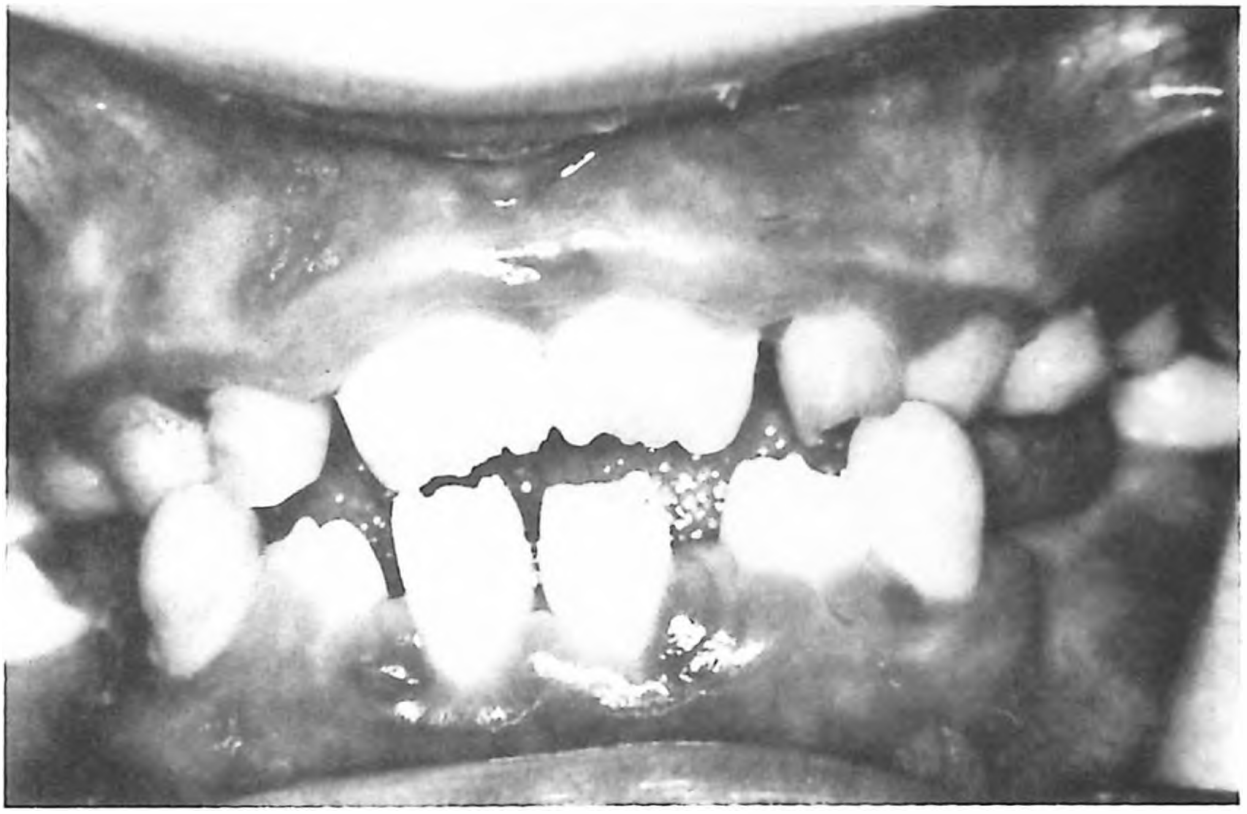

Fig. 1. Chronic inflammatory gingival enlargement localized to the anterior region.

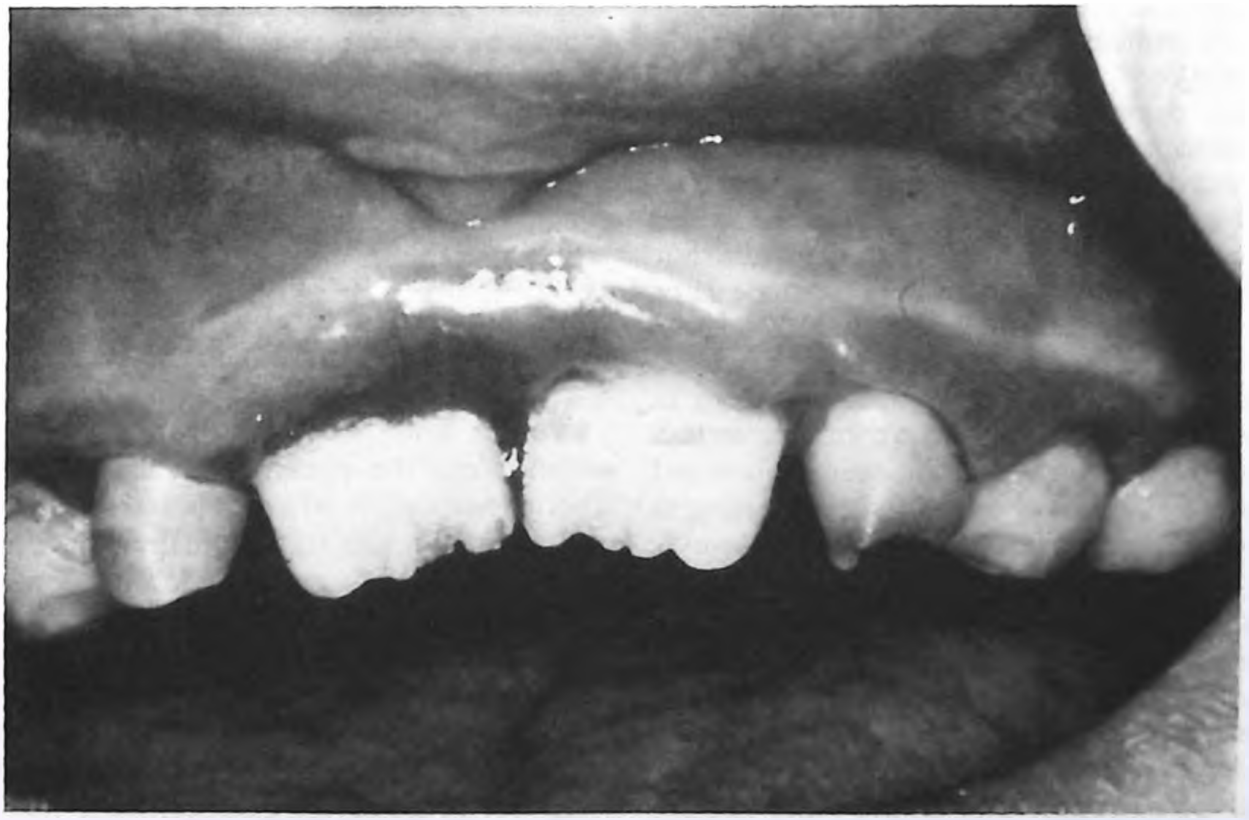

Fig. 2. Bleeding upon gentle manipulation due to the severe inflammation of the gingiva.

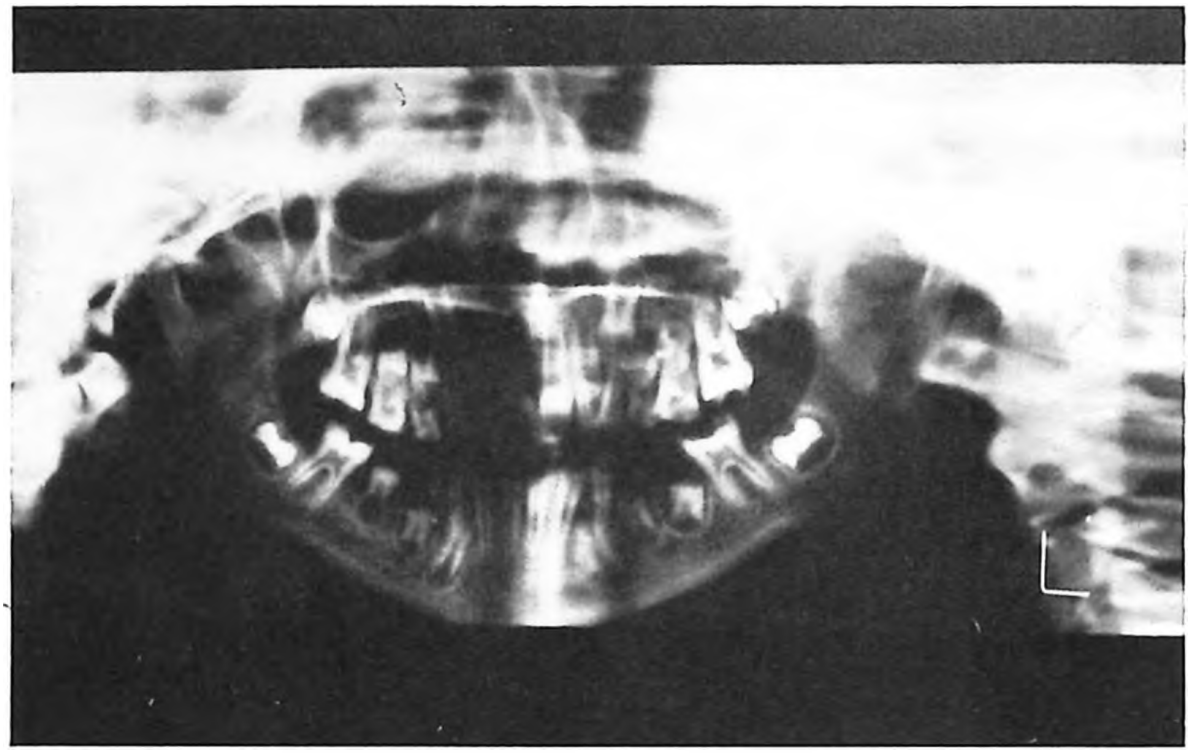

Fig. 3. Alveolar bone loss around the first molars.

Discussion

Chronic neutropenia of childhood is a group of disorders in which production of neutrophils is decreased or ineffective. The underlying mechanism has not been well identified yet. Autosomal dominant pattern of transmission has been reported. On the other hand, there are several reports indicating the absence of genetic transmission[9]. In the present case, Fanconi's anemia, dyskeratosis congenita, cartilage-hair hypoplasia, and Schwachman-Diamond syndrome were ruled out based on normal phenotype. The age at onset and the course of the disease as well as normaley of neutrophil counts were not consistent with congenital severe neutropenia or cyclic neutropenia. Therefore, she was diagnosed as chronic idiopathic neutropenia. In general, depressed resistance against microorganisms in chronic neutropenia and neutrophil dysfunction may disturb the interactions between the host and the microorganisms of bacterial plaque, leading to increased susceptibility to preiodontal disease[1-8].

Both gingivitis and chronic inflammatory gingival enlargement are caused by prolonged exposure to dental plaque[10]. Factors that favor plaque accumulation and retention include poor oral hygiene, abnormal relationships of adjacent teeth and opposing teeth, improper dental restorations, nasal obstruction and habits like mouth breathing[10]. Our patient had the habit of mouth breathing due to the nasal obstruction caused by recurrent upper respiratory tract infections and tonsillary hypertrophy. In addition, since she was in the mixed dentition period, abnormal teeth relations existed. Furthermore, her father noted that she resisted attempts to brush her teeth because of painful aphthous lesions. We think that the factors causing bacterial plaque accumulation may contribute to the clinical picture of generalized gingival inflammation and inflammatory gingival enlargement.

There are a few reports[2,3] describing periodontal findings in chronic idiopathic neutropenia, and they document that this type of neutropenia is characterized by oral symptoms of severe persistent gingivitis, recurrent occurrence of aphthous-type lesions and/or rapidly advancing alveolar bone destruction. As previously mentioned, the patient also had prepubertal periodontitis[11,12] which has its onset before 11 years of age in the primary or mixed dentition. Prepubertal periodontitis appears in Papillon-Lefèvre syndrome. Down syndrome. Chédiak-Higashi syndrome, hypophosphatasia, acute and subacute leukemia, leukocyte adhesion deficiency and neutropenias[11,12]. In our case, alveolar bone destruction, a characteristic sign of prepubertal periodontitis, was very limited at this stage, but she was included into

a periodontal follow-up program in order to prevent further alveolar bone loss which may result in exfoliation of the teeth.

In view of the previous reports[1-8,11,12] and our findings, we suggest that periodontal evaluation of patients exhibiting neutropenia is of utmost importance to avoid the destruction of supporting structures of the dentition. We also want to point out that gingival enlargement is not a hallmark of leukemia only.

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