Oral manifestations in congenital hypoplastic anemia (Diamond-Blackfan anemia): clinical report

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Abstract

Congenital hypoplastic anemia is a rare disease characterized by a normochromic, normocytic anemia, a deficiency of erythroblasts in the marrow, and normal leukocyte and platelet counts. The disease usually is treated successfully with corticosteroid therapy; however, some patients become refractory to corticosteroids and must receive washed, packed red blood cell transfusions. The management and unique dental findings of a 15-year-old patient with congenital hypoplastic anemia are described.

Clinical Report

A 15-year-old Caucasian male entered the dental clinic of Children’s Medical Center in Dallas, Texas, for routine dental care. Medical history at six weeks after birth revealed findings of dry scaly skin, ear deformity, anemia, and a webbed neck, diagnosed as spherocytosis. The family history was positive for spherocytosis on the maternal side. The diagnosis was changed to congenital hypoplastic anemia at eight weeks after birth and he was placed on corticosteroid therapy for nine years. At age 10, it was decided to discontinue the steroid therapy and to begin washed, packed red blood cell transfusions because of many complications including marked cushingoid characteristics, vertebral compression fractures, genu valgum, and severe osteoporosis.

Following removal from steroid therapy, there was a marked improvement in his cushingoid appearance, multiple carious lesions, and poor healing of recent extraction sites. The purpose of this clinical report is to present some unusual dental findings in a patient with this disease.
The patient had normal intelligence and no family history of congenital hypoplastic anemia. Current medications include two units of washed, packed red blood cells monthly and daily subcutaneous deferoxamine chelation therapy due to chronic iron overload. Hepatitis B surface antigen determination was negative and he was given hepatitis B antibody with his immunization in August, 1983. Recent laboratory results revealed a mild-to-moderate leukopenia and borderline thrombocytopenia which are not uncommon in patients with congenital hypoplastic anemia.

Past dental history revealed infrequent dental care at the dental clinic of Children’s Medical Center. Prior dental records revealed extraction of a supernumerary tooth adjacent to the mandibular right first premolar and also extraction of the maxillary and mandibular left third molars. Healing was reported to have occurred without incident. The maxillary central incisors were restored with mesiofacial composites at that time. There were no interim recall appointments until this current visit.

The patient received panoramic and intraoral radiographs. The radiographs were negative for dental caries, but revealed one third molar, and the unique finding of nearly total obliteration of the coronal pulp chambers of the erupted dentition (Figures 1-5). Intraoral examination revealed generalized marginal gingivitis secondary to poor oral hygiene, no evidence of bruxism, and a Class I molar relationship.

After consultation with the patient’s hematologist, an oral prophylaxis and topical fluoride treatment were performed.

Discussion

A dental patient with congenital hypoplastic anemia can present two areas of concern to the dentist. If treated with continual corticosteroid therapy, the patient may need additional corticosteroids prior to dental treatment and may be immunosuppressed. If the patient is refractory to corticosteroids and receives washed, packed red blood cell transfusions, dental treatment should be performed soon after the transfusions. Since a patient receiving transfusions may be a hepatitis B carrier, these patients should be tested for the presence of hepatitis prior to treatment. Dental personnel should, of course, take routine precautions associated with treating patients at high risk for hepatitis B. The dentist should consult and work closely with the patient’s hematologist.

The other report in the dental literature describing oral findings associated with congenital hypoplastic anemia makes no mention of unusual pulpal anatomy. Therefore this is the first case of coronal pulpal obliteration reported in association with this rare disease. Obliteration of pulp chambers is reported in association with three diseases: dentinogenesis imperfecta, osteogenesis imperfecta, and with a disorder similar to dentinogenesis imperfecta, dentinal dysplasia. However, teeth in these disorders exhibit abnormalities of crown and root morphology and tooth color, and excessive patterns of attrition. The patient in this report had teeth of normal color and crown morphology without excessive attrition or other features associated with these three disorders. Factors which can cause obliteration of pulp chambers are trauma, caries, and restorative procedures, though these factors affect only selected teeth. The finding of generalized coronal pulpal obliteration in this patient is a unique finding since no occlusal trauma or bruxism was present.