Unusual gingival swelling in a 4-year-old child

Anne-Laure Ejeil, DDS, PhD, Aude Thomas, DDS, Sébastien Mercier, DDS, and Nathan Moreau, DDS
Bretonneau Hospital, Paris, France, and Paris Descartes University, Sorbonne Paris Cité, France
(Oral Surg Oral Med Oral Pathol Oral Radiol 2014;118:627-631)

CLINICAL PRESENTATION

A 4-year-old female patient with gingival swelling was brought to the Department of Oral Surgery. The progressive swelling, present since birth, had been disturbing food intake. Chronic constipation (1 bowel movement every 3 to 4 days, treated with macrogol 3350) associated with intermittent rectal bleeding was also reported by her mother. The patient's rectal bleeding had been previously diagnosed as "hemorrhoids" by a gastroenterologist. Apart from these gastrointestinal manifestations, the patient appeared to be of normal weight and stature for her age (105 cm/15.7 kg).

Clinical extra-oral examination revealed cracked and swollen lips associated with a perioral exfoliating erythematous epidermal reaction (Figure 1).

Intra-oral examination revealed a generalized reddish gingival hypertrophy of firm consistency and granulomatous aspect involving the maxilla (Figure 2). Hyperplastic squamous linear lesions of the oral mucosa were also noted (Figure 3).

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of chronic granulomatous gingival hypertrophies should include sarcoidosis, granulomatosis with polyangiitis (formerly known as Wegener granulomatosis), orofacial granulomatosis, and Crohn disease.

Sarcoidosis is a chronic systemic disorder of unknown etiology characterized, in affected organs, by an accumulation of epithelioid granulomas without histopathologic aspects of caseation or presence of infectious agents.[1] Peak incidence is found between the ages of 20 and 40. The disease is more common among African Americans and Northern Europeans. Most reported pediatric cases have been described in patients aged 13 to 15. Sarcoidosis affects multiple organs, especially the lungs, lymph nodes, skin, and eyes. In the maxillofacial region salivary gland involvement is common. Clinically, most cases appear as hypertrophic or nodular lesions of firm consistency. Oral forms of sarcoidosis have mostly been described on the lips, hard and soft palate, buccal mucosa, gingiva, tongue, and tonsils. Oral involvement usually appears in patients with chronic multisystemic sarcoidosis. Patients may also present systemic manifestations such as fever, weight loss, and fatigue. Infants and children under the age of 5 usually present a triad of skin, joint, and eye involvement, without the typical lung disease.[1] Because several cases of oral sarcoidosis without systemic involvement have been described, sarcoidosis could not be ruled out of the differential. Nevertheless, gastrointestinal involvement is rare, occurring in about 3% of cases of sarcoidosis.

Granulomatosis with polyangiitis (GPA), formerly known as Wegener granulomatosis, is a vasculitis affecting small and medium caliber blood vessels. It results in nasal and pulmonary lesions or significant systemic involvement with renal, cardiac, neurologic, and skin lesions. The peak GPA incidences occur in the 30s and 40s, with a mean age of 41. It is rarely observed in children. Only 15% of cases occur in patients younger than 20, with a female predominance[2] and a higher prevalence among Caucasians (especially those from Northern Europe) compared with Asian, African, Afro-Caribbean, and African-American populations. Constitutional symptoms of active granulomatosis with polyangiitis are general malaise, myalgia, arthralgia, anorexia, weight loss, and pyrexia. Renal and pulmonary diseases are common findings at time of diagnosis. Cytoplasmic antineutrophil cytoplasmic antibodies (cANCA) and perinuclear anti neutrophil cytoplasmic antibodies (pANCA)-ANCA are considered sensitive and specific markers for GPA.[3] Oral involvement classically presents as hyperplastic granular gingivitis.[4,5] Atypical oral ulcerations have also been observed.[6] Diagnosis is based on a combination of positive ANCA serology and histologic evidence of necrotizing vasculitis, necrotizing glomerulonephritis or granulomatous inflammation from a relevant organ biopsy, such as skin, lung, or kidney. Our patient did not present any signs of systemic involvement suggestive of vasculitis.

Statement of Clinical Relevance

Oral manifestations of Crohn disease may precede the onset of intestinal disease in up to 60% of cases. Knowledge and detection of such manifestations may help in early diagnosis and better management of this disease.

Bretonneau Hospital, Department of Oral Surgery, Paris, France, and Paris Descartes University, Sorbonne Paris Cité, France.
Received for publication Feb 9, 2014; returned for revision May 21, 2014; accepted for publication May 30, 2014.

2212-4403/$ - see front matter
http://dx.doi.org/10.1016/j.oooo.2014.05.024

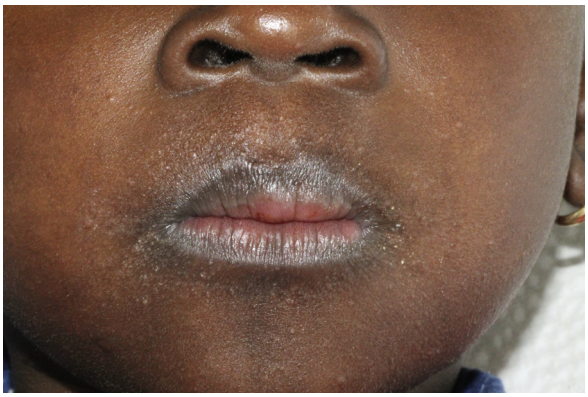

Fig. 1. Significant swelling and fissuring involving the upper lip.

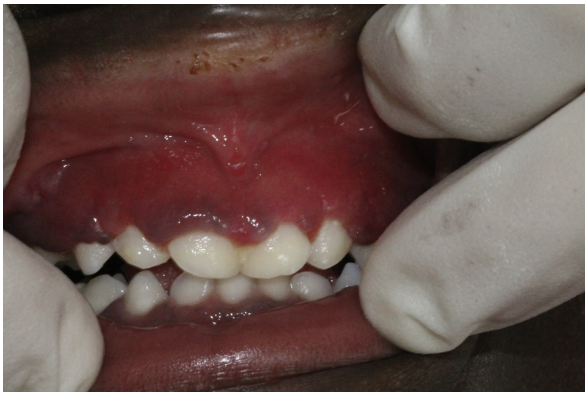

Fig. 2. Intra-oral photography showing an erythematous and swollen gingiva.

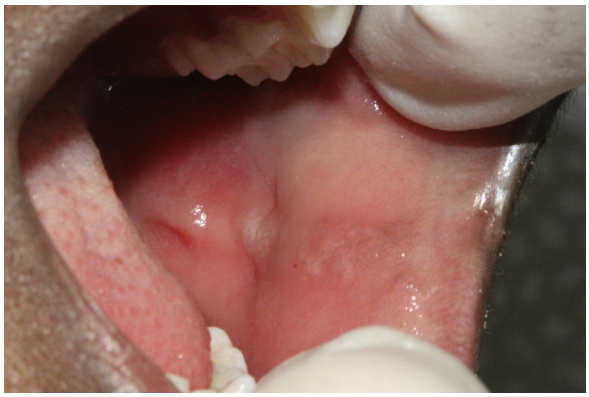

Fig. 3. Linear hyperplasic lesions of the oral mucosa.

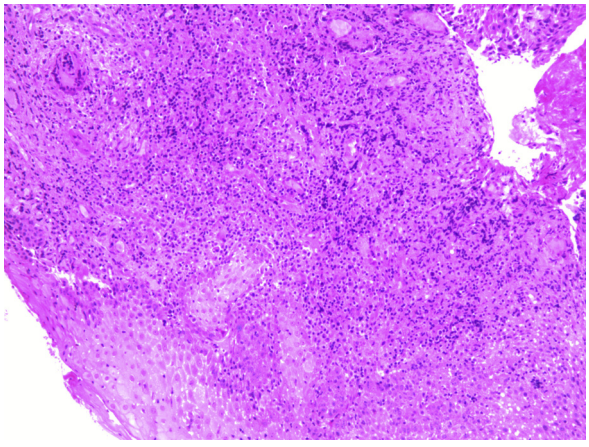

Fig. 4. Gingival biopsy showing giant cell granulomas without necrosis (hematoxylin-eosin, magnification ×100; courtesy Tolbiac Laboratory, Dr. F. Le Pelletier).

Orofacial granulomatosis (OFG), which includes Melkersson-Rosenthal syndrome (MRS) and Miescher cheilitis granulomatosa, is a group of chronic inflammatory diseases characterized by noncaseating granulomatous inflammation and affecting oral and maxillofacial soft tissues. MRS has been described as a syndrome of unknown etiology characterized by the triad of persistent lip or facial swelling, recurrent facial paralysis, and fissured tongue. When faced with either facial swelling or cheilitis granulomatosa without the other 2 components of the triad, clinical manifestations should be labeled orofacial granulomatosis. A diagnosis of OFG is made through histopathologic identification of noncaseating granulomas. MRS was ruled out based on the absence of facial paralysis and a tongue of normal clinical aspect, but we could not rule out OFG.

Crohn disease (CD) is a chronic, idiopathic, inflammatory, granulomatous disease of unknown etiology involving the gastrointestinal tract. Common symptoms include abdominal pain, diarrhea, fatigue, fever, gastrointestinal bleeding, and weight loss.[7] Oral lesions have been described in patients with intestinal CD.[8] These include swelling of the lips and buccal mucosa, gingival hyperplasia, cobblestone appearance of the oral mucosa, mucogingivitis, deep linear ulcerations, and mucosal tags. Colonoscopy, including ileoscopy and biopsy, are often useful in the diagnosis of ileo-colic Crohn disease.[7] Diagnosis of CD is made based on histopathologic identification of noncaseating granulomas in association with gastrointestinal manifestations and serologic disturbances. Infectious granulomatous conditions (such as mycosis or tuberculosis) must be excluded by use of special staining methods and cultures.

In this case, we favored a granulomatous lesion, not otherwise specified.

DIAGNOSIS AND MANAGEMENT

Considering the absence of definitive clinical manifestations, an incisional biopsy of the gingiva was performed. Hematoxylin and eosin staining revealed a non-necrotic giant cell granuloma compatible with Crohn disease and with sarcoidosis (Figure 4). Periodic acid−Schiff, Grocott, and Ziehl-Neelsen stains were then applied to biopsy specimens to rule out fungal and