Strawberry gingivitis as the first manifestation of granulomatosis with polyangiitis

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A 49-year-old previously healthy male patient was referred to the Institute of Dentistry, Jagiellonian University Medical College, Kraków, Poland, because of bleeding hypertrophic lesions on his gum, which developed a few weeks earlier. In the past 6 months, he had lost 7 kg without dieting, and he complained of persistent fatigue. One month before the oral manifestations, he had had several episodes of epistaxis and developed significant persistent hearing loss and tinnitus on the right side. The laboratory tests revealed lymphopenia (0.8 x 10⁹/µl; 11% of the total leucocyte count of 7.26 x 10⁹/l), an increased level of C-reactive protein (90 mg/l), and discrete proteinuria (0.09 g/l) with active urine sediment presenting as 5 to 8 red blood cells per high power field. Other kidney function parameters were within the normal range. Intraoral examination showed circumscribed, erythematous gingival enlargement typical for “strawberry-like” gingivitis. The swollen, reddened, and granular pathological lesions were located mainly on the gingiva and mucosa in the region of the right maxillary first premolar and second incisor, and the left incisors and canine (figure 1A). In the mandible, the lesions were less extensive and were located in the region of the left first premolar, canine, and incisors, and on the right side, in the second incisor (figure 1B). Since these lesions are most often found in granulomatosis with polyangiitis (GPA; formerly known as Wegener’s granulomatosis), the patient was referred to the autoimmune diseases department for further tests. No tissue biopsy was taken owing to the history of intense bleeding. Further laboratory tests revealed positive results for antineutrophil cytoplasmic antibodies (ANCA; 1:40 titer) directed against proteinase 3 (PR3, cANCA; 72 RU/ml). A computed tomography scan showed inflammatory masses in the paranasal sinuses and middle ear on the right side, and multiple nodules in both lungs. Significant conductive hearing impairment was confirmed using pure tone audiometry, while pulmonary function tests remained normal. Diagnosis of GPA was established, and treatment was initiated with methylprednisolone and cyclophosphamide (CTX) pulses. After 10 weeks of treatment (cumulative CTX dose of 4 g), the patient’s general condition improved, gingival lesions completely resolved, and no further bleeding episodes from the nose and gums were reported.
GPA is a rare inflammatory disease of the small vessels, characterized by the presence of ANCA. The disease most commonly manifests in the paranasal sinuses, respiratory tract, and kidneys. Massive renal injury or prominent lung involvement as the first disease manifestations tend to progress rapidly leading usually to rapid diagnosis. More challenging, from the primary care or outpatient perspective, are nonspecific musculoskeletal, skin, and mucosal manifestations or chronic refractory sinusitis. Oral cavity lesions in GPA may present as nonspecific aphthae or erosions, or in rare cases, as purple gingival hypertrophy (sometimes with petechial hemorrhages) known as strawberry gingivitis. The latter is an almost pathognomonic mucosal symptom of vasculitis. In this case, a lack of prominent renal involvement and respiratory symptoms delayed proper diagnosis. Once strawberry gingivitis was recognized as a possible sign of GPA, effective treatment could be initiated without delay.

REFERENCES