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Oral granulomatosis with polyangiitis

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Granulomatosis with polyangiitis (GPA), formerly named Wegener's granulomatosis is an autoimmune, rare disease characteristic for systemic vasculitis.[1] Currently, the etiology and pathogenesis of GPA are still unknown. This disease has a wide age range, with a mean age of 41 years and 85% of cases older than 19 years of age. There is no prominent gender predilection.[2] Classic GPA initially involves the upper and lower respiratory tract with symptoms such as cough, hemoptysis, and pleuritis.[3] Oral lesion has been found in 6–13% of patients, and oral involvement as the initial presentation of GPA is only identified in 2% of cases.[3] In this study, we presented a case of GPA who was diagnosed from the oral signs.

The 44-year-old female patient felt pain and swelling of the pan-oral gingiva for one month. She had been to a local dental clinic for treatment, and the dentist suggested her to visit our OPD for further examination. Upon oral examination, hyperplastic, hemorrhagic friable gingiva was present at her labial side of upper anterior teeth (Fig. 1A). Multiple ulcerative granulomatous lesions of pan-oral buccal and lingual gingivae were also noted (Fig. 1B and C). The periapical radiographs revealed moderate horizontal bony defects of the alveolar process (Fig. 1D). Due to the atypical gingival swelling resembled a characteristic oral manifestation of GPA known as strawberry gingivitis, an oral involvement of GPA was highly suspected. Medical checkup and incisional biopsy for gingival lesions were arranged. She had elevated white blood cells (13,830 cells/μL (normal range: 4140–10520 cells/μL)) and C-reactive protein level (94.98 mg/L (normal range: $<$ 5 mg/L)). The PR3-ANCA (proteinase 3-antineutrophil cytoplasm antibody) was 7.8 IU/mL (normal range: $<$ 2.0 IU/mL). The urinary screening revealed hematuria and proteinuria. The histopathological examination showed an ill-defined, necrotizing granulomatous lesion. The covering epithelium displayed pseudoepitheliomatous hyperplasia (Fig. 1E). The supporting stroma contained proliferation of vascular channels and a heavy inflammatory infiltrate consisting of neutrophils, lymphocytes, and multinucleated giant cells (Fig. 1F and G). Some subepithelial abscesses and extravasation of red blood cells were also seen. The histopathological features were consistent with a GPA. Then she was admitted to our rheumatology ward for further treatment.

The orofacial manifestations of GPA include strawberry gingivitis, ulceration, facial paralysis, labial mucosal nodules, sinusitis of the maxillary sinus, and sialadenitis of salivary glands.[4] The diagnosis of a GPA requires more than two diagnostic criteria proposed by the American College of Rheumatology, which include (1) oral ulcers or nasal discharge, (2) nodules, fixed infiltrates, or cavities on a chest radiograph, (3) abnormal urinary sediment (red blood cell cast or $>$ 5 red blood cells per high power field), (4) granulomatous inflammation upon biopsy.[3] Despite the diagnosis of a GAP may be challenging, and the oral presentation of GPA is very uncommon. Dentists play an important role in early diagnosis, especially in

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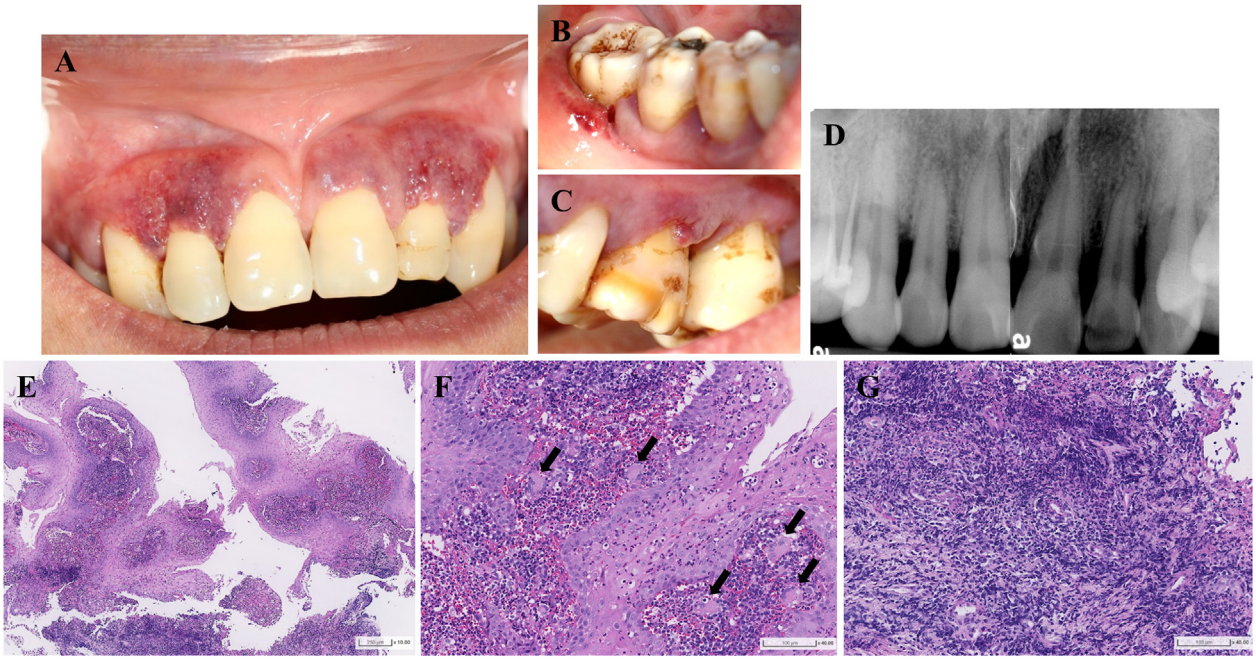

Figure 1 Clinical, radiographic, and microscopic photographs of the current case of GPA with oral involvement. Hyperplastic, hemorrhagic friable gingiva was present at labial gingiva of tooth 13 to 23 (A). Multiple ulcerative granulomatous lesions were identified at pan-oral buccal and lingual gingivae (B and C). The periapical radiographs of upper anterior teeth revealed moderate horizontal bony destruction of the alveolar process (D). The histopathological examination showed an ill-defined, necrotizing granulomatous lesion with papillary surface. The covering epithelium displayed pseudoepitheliomatous hyperplasia (E). Numerous multinucleated giant cells (arrow) were present in the granulation tissue (F). Proliferation of vascular channels and a heavy inflammatory infiltrate consisting of neutrophils and lymphocytes (G). (Hematoxylin and eosin stain; original magnification; E, 10×; F and G, 40 ×).

cases with oral signs prior to pulmonary and renal involvement.[5]

Declaration of competing interest

The authors have no conflicts of interest relevant to this article.

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