

Strawberry gingivitis as the first sign of Wegener's granulomatosis: a case report



CrossMark

Yongky Tamigoes,* Riani Setiadhi*

Abstract

Objective: To describe that Wegener's granulomatosis (WG) is an autoimmune multisystem disease with unknown etiology, characterized by the classic triad of necrotizing granulomas affecting the upper and lower respiratory tracts, disseminated vasculitis and glomerulonephritis. WG can affect any organ including kidneys, eyes or other organs. Oral involvement, like strawberry gingivitis, is an important sign of this disease even though it is rare.

Methods: A 19 years old female was referred to oral medicine clinic with the chief complaint swelling on the gingiva which was increasing widespread. Since one month before the gingival enlargement did not respond to antibiotic. Intraoral examination, showed gingival enlargement at the upper anterior, palate, and first left mandibular molar. The swollen tissue was characterized by its

granularity, short bulbous projections and bright red colour. The gingival appearance mimicking an overripe strawberry and extra oral examination showed cutaneous vasculitis at the hands and feet. Laboratory evaluation presented erythrocyte sedimentation rate, CRP and eGFR (modified MORD) were elevated but the urinalysis was within normal ranges. Based on the anamnesis, clinical and laboratory examination, WG diagnosis was made. The patient was treated with oral corticosteroids and chlorhexidine gluconate 0.1% mouthwash.

Results: The gingival inflammation improved after one month therapy and the patient is still being followed-up at regularly time.

Conclusions: Early detection of oral manifestation of WG is important for dentists, especially oral medicine specialist, in order to achieve adequate treatment and preventing systemic tissue or organ damage.

Keywords: Oral manifestation, Strawberry gingivitis, Wegener granulomatosis

Cite this Article: Tamigoes Y, Setiadhi R. 2019. Strawberry gingivitis as the first sign of Wegener's granulomatosis: a case report. *Journal of Dentomaxillofacial Science* 4(1): 49-52. DOI: [10.15562/jdmfs.v4i1.741](https://doi.org/10.15562/jdmfs.v4i1.741)

Department of Oral Medicine,
Faculty of Dentistry, Universitas
Padjadjaran, Bandung, Indonesia

Introduction

Wegener's Granulomatosis (WG) is an autoimmune multisystem disease with unknown etiology.^{1,2} A number of exogenous factor have been suggested to be the aetiological relevance because microbial exogenous factors may possibly rising the disease expression. The microbial agent probably causes non-specific activation of the immune system, resulting in elevation of cytokine levels in the presence of Antineutrophil Cytoplasmic Antibody (ANCA) and leading to cell destruction.³⁻⁵

There are two types of Wegener granulomatosis i.e generalized and limited: a generalized form in which have classic triad of granulomatosis namely vasculitis, glomerulonephritis and airway involvement. Limited forms of the respiratory tract and the musculo-skeletal system without renal involvement. The limited form of Wegener Granulomatosis can develop into a common form of WG where in the course of the disease there is renal involvement.⁶

One of the rare but important signs of this disease is oral involvement, occurring generally in 6-13% of patients, however oral involvement as the primary manifestation of the disease, occurs only in 5-6% of cases and may be the first diseases symptom. The most common manifestation of WG is strawberry gingivitis which is a pathognomic

symptom of this disease.⁷ Gingival involvement can be seen as multiple, small, bulbous and fragile granular hyperplasia and bleeding bumps. These red bumps are responsible for strawberry-like appearance in this disease.⁸

Only in a few studies, WG diagnosis was primarily approved based on oral symptoms and gingival involvement. In this paper, a case of WG that was primarily diagnosed on the basis of oral manifestations is reported.

Case Report

A 19 years old female was referred to the oral medicine specialist clinic at Dr Hasan Sadikin Dental Hospital with the chief complaint swelling on the gingiva which was increasing widespread. The patient was first diagnosed as hyperplastic gingivitis by a dentist at BA hospital and was treated with antibiotic since a month ago but didn't improved. Extra oral examination showed cutaneous vasculitis at the hands and feet.

Intraoral examination, showed gingival enlargement on the upper anterior tooth to posterior tooth, palatal gingivae, and first left mandibular molar [figure 1](#). The swollen tissue was characterized by its granularity, short bulbous projections and bright

*Corresponding to:
Yongky Tamigoes, Riani Setiadhi
Department of Oral Medicine,
Faculty of Dentistry, Universitas
Padjadjaran, Bandung, Indonesia
yongkytamigoes@gmail.com;
riani.setiadhi@fkg.unpad.ac.id

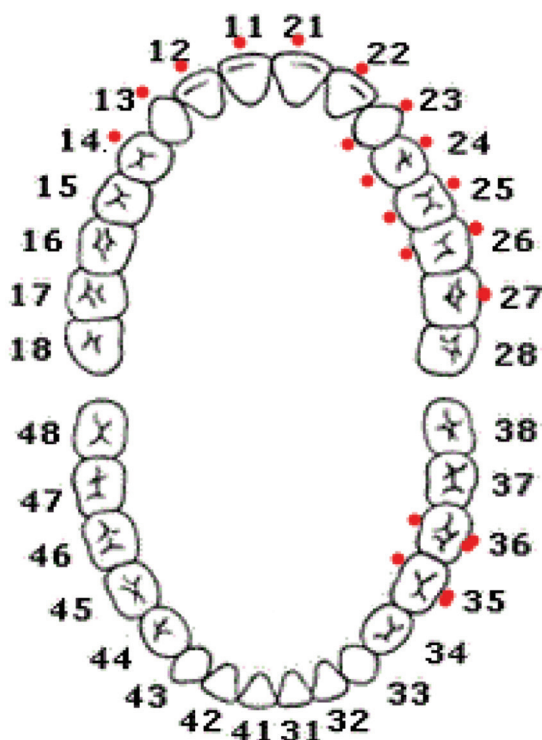


Figure 1 The patient's diagram gingival involvement



Figure 2 Patient's gingivae at the first visit

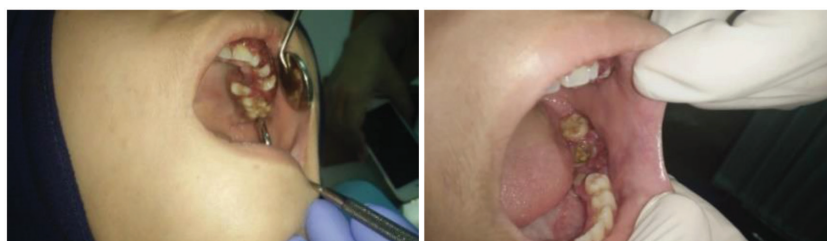


Figure 3 Patient's gingivae at second visit follow-up after one month treatment



Figure 4 Patient's gingivae at 2 months after first visit

red colour. The gingival appearance mimicking an overripe strawberry [figure 2](#). Laboratory examinations in patients can be seen in [table 1](#).

Based on the anamnesis, clinical and laboratory examination, diagnosis of WG was made and her treatment was started. The patient was referred to a rheumatologist for investigations and treatment but she only once visited the rheumatologist therefore the treatment was not finished yet. At the first visit, she received remission-induction treatment with oral corticosteroids and chlorhexidine gluconate 0.1% mouth wash. One month later, the gingival inflammation had greatly improved and the patient has been follow-up continuously at regular time [figure 3](#).

After 2 months from the first visit, oral lesions had improved and the gingiva looked healthy [figure 4](#). Four months later, the patient had epistaxis, cough with phlegm, joint pain and low vision but there were no specific oral problems [figure 5](#).

Discussion

The first and only symptom of the patient in this case report which showed that it was Wegener's granulomatosis were gingival exophytic growth and strawberry-like enlargements with petechiae. If the classic triad of necrotizing granulomatous lesions of the respiratory tract, generalized vasculitis and necrotizing glomerulonephritis are absent, the diagnosis of WG is challenging.¹ It has been suggested that oral mucosal and cutaneous vasculitis findings are valuable diagnostic aids in WG.⁹

Almouhawis et al.⁷ stated that WG has a wide range of non-specific characteristics as it can affect multiple systems, making early diagnosis difficult. The upper respiratory tract, lungs and kidneys are most commonly involved.^{10,11} However, other sites that could be involved, including the central nervous system, skin, salivary glands, oral cavity, eyes, heart and musculo-skeletal involvement.¹² The oral lesions may manifest either as mucosal ulcer and as gingival hyperplasia with classical "strawberry gingivitis."

Gingival enlargement or gingival hyperplasia is usually an adverse effect of drugs such as phenytoin, cyclosporine, an immunosuppressive drug and calcium channel antagonist. In phenytoin and calcium channel blockers induced gingival overgrowth, the gingiva usually appears pink, firm to spongy and rubbery. Our patient did not use any drugs known to cause gingival overgrowth. Clinical features of gingival enlargement can also be seen in crohn's disease. In crohn's disease, the gingiva is pink, toned and almost rough in consistency, with a very pinched



Figure 5 Patient's gingivae at 4 months after first visit

Table 1 Result of Laboratory Investigations

Test	Result	Normal Value
Blood count		
Hemoglobin	12.7	12-16 g/dl
Hematocrit	38.2	35-47 %
leukosit	7.93	4.40-11.30 $\times 10^3$ /uL
RBC (red blood cell)	4.51	3.60 – 5.80 million/uL
Platelets count	406.000	150.000-450.000/uL
MCV	84.7	80-100 fL
MCH	28.2	28.2 pg
MCHC	33.2	32-26%
Erythrocyte Sedimentation Rate (ESR)	66	0-25 mm/hour
C-reactive Protein (CRP)	1.74	< 0.3 mg/dL
eGFR (modified MORD)	170.5	104-148 mL/min/1.73m ²
Creatinine	0.6	0.6-1.1 mg/dL
Aspartate Transaminase (AST),	12	15-37 U/L
Alanine aminotransferase (ALT)	11	14-59 UL
urinalysis	Normal	
Imunoserology		0-387 IU/mL
Ig E	275.6	
ANA	Negative	
ANCA	Negative	

surface.¹³ However, in our patients the clinical feature showed gingival enlargement mimicking an overripe strawberry, therefore the diagnosis of crohn's disease was excluded.

In the case presented, diagnosis of WG was made supported by the results of laboratory tests showing an increase in ESR, CRP. Although, the results of the cANCA examination were negative in this patient. The negative cANCA results do not exclude the diagnosis of WG because the cANCA examination results are not the diagnostic criteria. However, cANCA results remain to be used to support the diagnosis of WG, because the positive cANCA results from 80-90% of patients with WG and 55-66% are positive in the WG limited. The results of cANCA also illustrate the disease's activity. In patients with limited clinical manifestations or

mild symptoms, cANCA can be negative in more than 40% of cases.¹⁴

After careful exclusion of the above-mentioned lesions by appropriate systemic evaluation, laboratory investigation and referring the previous reported cases with similar clinical appearance, treatment was initiated with prednisone. Hoffman et al.¹⁵ stated that the standard therapy of initial WG consisted of glucocorticoids or cyclophosphamide, or a combination of both. The patient was treated with oral prednisone 30 mg/day and chlorhexidine gluconate 0.1 % mouthwash. The provision of this prednisone in accordance with standard therapy of WG as stated by Hoffman and given chlorhexidine gluconate 0.1 % mouthwash to maintain the oral hygiene of this patient.^{15,16} One month later, the gingival inflammation had greatly improved. Once remission is achieved, prednisone is usually tapering of gradually. At the two months follow up, oral lesions had improved, the gingiva looked healthy and prednisone was tapering of gradually until alternate days at the third month. However, at the four months follow up, the patient had epistaxis, cough with phlegm, joint pain and low vision. This condition occurred because the patient only once visited the rheumatologist so that patient was not treated thoroughly.

Identification, early diagnosis and prompt treatment of strawberry gingiva are important in order to get a better prognosis. The appearance of strawberry gingiva is unique, distinct and pathognomonic, which makes it easily identifiable by an expert dental practitioner especially oral medicine specialist. Once the diagnosis has been made, medical treatment can be instituted promptly, which will dramatically improve the prognosis for patients with this serious, potentially fatal disorder. In 82% of cases that undiagnosed or untreated, the patient will not survive more than a year.⁶ However, with early initiation of appropriate treatment, up to 75% of patients with Wegener granulomatosis can expect significant or even complete remission.¹⁷

Conclusion

Early detection of oral manifestation of WG is important for dentists, especially oral medicine specialist, in order to achieve adequate treatment and preventing further systemic tissue or organ damage.

Acknowledgment

The authors would like to thank The Director of General Hospital Dr. Hasan Sadikin and The Head of SMF Dental Hospital Dr. Hasan Sadikin for the opportunity given to us to treat the patient.

Conflict of Interest

The authors report no conflict of interest.

References

1. Aravena V, Beltrán V, Fuentes R, et al. Gingival hyperplasia being the first sign of Wegener's granulomatosis. *Int J Clin Exp Med* 2014;7: 2373-2376.
2. Rashed M, Mozaffari PM, Javadzadeh A, et al. Wegener granulomatosis with oral involvement as primary manifestation: a case study. *JDMT* 2017;6: 89-96.
3. Yi ES, Colby TV. Wegener's granulomatosis. *Semin Diagn Pathol* 2001;18: 34-46.
4. George J, Levy Y, Kallenberg CG, et al. Infections and Wegener's granulomatosis cause and effect relationship. *QJM* 1997;90: 367-373.
5. Grahne B, Zechner G. Wegener's granulomatosis. *Acta Otolaryngol* 1966;62: 362-372.
6. Siar CH, Yeo KBKNHN, Tsujigiwa H, et al. Strawberry gingivitis as the first presenting sign of Wegener's granulomatosis: report of a case. *Eur J Med Res* 2011;16: 331-334.
7. Almouhawis HA, Leao JC, Fedele S, et al. Wegener's granulomatosis: a review of clinical features and an update in diagnosis and treatment. *J Oral Pathol Med* 2013;42: 507-516.
8. Manchanda Y, Tejasvi T, Handa R, et al. Strawberry gingiva: a distinctive sign in Wegener's granulomatosis. *J Am Acad Dermatol* 2003;49: 335-337.
9. Marzano AV, Balice Y, Papini M, et al. Localized Wegener's granulomatosis. *J Eur Acad Dermatol Venereol* 2011;25: 1466-1470.
10. Lourenço SV, Nico MMS. Strawberry gingivitis: An isolated manifestation of Wegener's granulomatosis. *Acta Derm Venereol* 2006;86: 90-91.
11. Comarmond C, Cacoub P. Granulomatosis with polyangiitis (Wegener): clinical aspects and treatment. *Autoimmun Rev* 2010;13: 1121-1125.
12. Guidelli GM, Tenti S, Pascarelli NA, et al. Granulomatosis with polyangiitis and intravenous immunoglobulins: a case series and review of the literature. *Autoimmun Rev* 2015;14: 659-664.
13. Agrawal AA. Gingival enlargements: differential diagnosis and review of literature. *World J Clin Cases* 2015;3: 779.
14. Hiz F, Cinar M, Karagol T, et al. Wegener's granulomatosis: case report. *Internet J Neurol* 2007; 6.
15. Hoffman GS. Wegener's granulomatosis. *Curr Opin Rheumatol* 1993;5: 11-17.
16. Muryah AE, Sufiawati I. Successful treatment of herpes simplex-associated erythema multiforme with a combination of acyclovir and prednisone. *J Dentomaxillofac Sci* 2017;2: 191-193.
17. Cotch MF, Hoffman GS, Yerg DE, et al. The epidemiology of Wegener's granulomatosis. Estimates of the five year period prevalence, annual mortality and geographic disease distribution from population-based data sources. *Arthritis Rheum* 1996;39: 87-92.



This work is licensed under a Creative Commons Attribution