CASE STUDY

Wegener’s Granulomatosis With Oral Mucosal Involvement as First Manifestation

Granulomatosis de Wegener con afectación de la mucosa oral como primera manifestación

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Introduction

Wegener’s granulomatosis (WG) is an autoimmune disease, which particularly affects the upper respiratory pathways, lungs and kidney. Oral mucosal involvement presents in around 5%–10% of cases and may be the first disease symptom. Predominant manifestation is granulomatous gingivitis erythematous papules; mucosal necrosis and non-specific ulcers with or without impact on adjacent structures. Clinically speaking, the most characteristic lesion presents as a gingival hyperplasia of the gum, with hyperaemia and petechias on its surface which bleed when touched. Due to its appearance, it has been called “Strawberry gingiva”. The following is a clinical case in which the granulomatous strawberry gingivitis was the first sign of WG.

Clinical Case

A gypsy woman aged 44 was referred from another hospital’s A&E department following examination by an otolaryngologist, after presentation with a two-month history of pain and swelling across the upper right dental arch, accompanied by fever during the previous 2 weeks. Prior to this period, a dental implant in the upper right premolar with no prosthesis had been fitted. Medical treatment had been received with diverse antibiotics and mouthwashes but no improvement had been made.

Anamnesis was with no findings of interest. She was on occasional smoker of less than half a packet of cigarettes per day.

Granular and friable gingival formations were discovered on oral examination. These were deep red in colour in both dental arches (Fig. 1) and bled when touched. The orthopantomograph showed an implant in position 15 with no signs of bone resorption or rarefaction of adjacent bone. All other complementary examinations’ findings were normal.

On hospital admission a computerised tomography (CT) of the face and neck was requested. This revealed subcutaneous cellular tissue inflammation of the upper right dental arch with several necrotic-cystic lesions and swelling of the right apical pleura. Results of analysis showed leucocytosis, increased VSG=172, perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA) <3.10 (negative), and perinuclear anti-neutrophil cytoplasmic antibodies (c-ANCA) of 5.10 (mildly positive).

On suspicion of an associated pleural effusion, the study was completed with a CT scan of the thorax, which showed total condensation on the upper right lobe.

Infectious/pulmonary neoplasm processes were ruled out by CT scans of the thorax, abdomen and pelvis, Mantoux, bronchoalveolar lavage (BAL), bronchoalveolar aspirate (BAS), and fibrobronoscopy, all of which resulted normal.

On 2 occasions samples were taken of the affected gum. The first showed chronic non-specific inflammation.
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The second, coinciding with an c-ANCA blood peak above 30 pg/ml, showed inflammation, necrosis, granulomas and multinucleated giant cells compatible with WG (Fig. 2). Treatment was initiated with cyclophosphamide and prednisone, which resulted in clinical improvement of the patient after four days. After one week, the granulomatous lesions had almost disappeared from the gum. The patient was periodically monitored by our hospital’s rheumatology service.

Discussion

Although the case we present does not comply with the classical triad relating upper respiratory tract mucous membrane, lower respiratory tract, and kidneys, the gingival affection described is an alarm signal alerting possible WG. The discovery of pseudoepitheliomatose hyperplasm from an attached gum biopsy, with multinucleate giant cells and microabscesses confirmed diagnosis of WG. It has even been described that classical criteria such as vasculitis, granulomas, and necrosis are notably absent in gum biopsies.

Differential diagnosis of this type of gingival alteration should be made with local and well defined systemic entities (Table 1). In all previously mentioned cases, the key resides in the anti-neutrophil cytoplasmic antibodies titre (c-ANCA), which is negative in said conditions and positive in WG. However, clinical suspicion together with compatible histopathological study is more sensitive in diagnosis than c-ANCA levels, as they may fluctuate. Drug-induced gingivitis is usually slim and fibrotic, and does not fit the description of this clinical case. There is no correlation of c-ANCA values with disease activity.

Treatment with cyclophosphamide and prednisone improved the patient’s condition and, as observed, led to the remission of the oral lesions after a few days.

Table 1 Differential Diagnosis of the Granulomatous Gingivitis.

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<thead>
<tr>
<th>Local medical condition</th>
<th>Systemic medical condition</th>
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<tr>
<td>Oral manifestation of HIV</td>
<td>Crohn’s disease</td>
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<tr>
<td>Tuberculosis</td>
<td>Wegener’s granulomatosis</td>
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<td>Leishmaniasis</td>
<td>Sarcoidosis</td>
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<td>Syphilis</td>
<td>Midline granuloma</td>
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<td>Actinomycosis</td>
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<td>Histoplasmosis</td>
<td>Langerhans cell histiocytosis</td>
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<td>Coccidioidomycosis</td>
<td>Drug-induced gingivitis</td>
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<td>Blastomycosis</td>
<td>Phenytin</td>
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<td>Mucormycosis</td>
<td>Cyclosporine</td>
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<td>Aspergillosis</td>
<td>Verapamil</td>
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Figure 1 Mucosal gingival lesions.

Figure 2 Histological study. Haematoxilin/eosin. Right (4×): histiocytic granuloma with palisade nuceli. Left (10×): necrotising vasculitis.
Conclusion

On suspicion of a case of WG, following presentation of non-specific gingivitis erythematous papules, confirmation requires c-ANCA in blood and repeated biopsies.

Conflict of Interest

The authors declare no conflict of interest.

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References