Oral Surgery ISSN 1752-2471

CASE REPORT

Strawberry gums in Wegener's granulomatosis: a rare presentation

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Key words:
medicine, oral, orofacial

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Accepted: 7 December 2007

doi:10.1111/j.1752-248x.2007.00012.x

Abstract

Wegener's granulomatosis is a multisystem disease characterised by necrotising granulomatosis of the upper and lower respiratory tracts, disseminated vasculitis and glomerulonephritis. However, the clinical manifestations and organs involved in the disease vary widely. This case report documents the importance of considering Wegener's granulomatosis in patients with unique oral lesions. Failure to recognise clinical lesions can result in delayed diagnosis and treatment, with potentially fatal results.

Case report

A 4-year-old boy presented on a consultant clinic in the department of oral and maxillofacial surgery, at a local hospital with a lesion on his left buccal gingivae which was apparently growing in size. He also complained of painful bleeding gums and oral ulcers since 4 months ago. The child was the first of the twins born at 36 weeks by caesarean section. His development history was unremarkable. In the past he had history of glue ear which had affected his hearing.

Examination revealed florid erythematous gingival hyperplasia with desquamation in the palatal aspect. He also had some nasal obstruction with mucus in the nose.

The gingivae were biopsied under GA and a number of haematological tests were also conducted. The biopsy showed non-specific granulomatous inflammation with epithelioid granulomas, which confirmed the clinical suspicion of Wegener's granulomatosis (WG). The child was prescribed co-trimoxazole and prednisolone for 2 weeks and improved with the initial treatment but there was still evidence of swelling on the labial aspect of the gingivae with erythema and hyperplasia over the interdental papillae.

He was urgently referred to a consultant paediatrician in a local university hospital to evaluate and rule out possible multisystem organ involvement, although he did not present with any systemic complaints and was symptom free. After treatment with prednisolone for 3 months the gingival hyperplasia resolved completely. He was on maintenance dose of 1 mg of prednisolone for 1 year and has been aymptomatic.

Differential diagnosis

Given the history, the differential diagnosis includes aetiologies such as tuberculosis, aspergillosis or sarcoidosis and even Crohn's disease. Other entities, which could be considered, include leukaemia, Churg-Strauss disease, polyarteritis nodosa and scurvy.

Investigations

Diagnosis of Wegener's is predominantly based on clinical features, a raised PR3-ANCA [proteinase 3 antineutrophil cytoplasmic antibodies, formally called cytoplasmic-antineutrophil cytoplasmic antibodies (c-ANCA)] and histopathological confirmation[1]. In mild cases of Wegener's, as in this one, PR3-ANCA can be

negative in up to 40% of individuals. Although testing for PR3-ANCA remains controversial as a clinical diagnostic tool, it is often the diagnostic feature in patients with non-specific biopsies. Other investigations include renal function tests, urinalysis (dipstick/proteinuria/haematuria), full blood count including erythrocyte sedimentation rate (ESR) and chest X-ray.

In this case, the laboratory investigations revealed markedly elevated platelets (735×10^9/L), a raised ESR (30 mm/h) and microcytic anaemia, although the c-reactive protein – 6 mg/L was within normal limits. A chest X-ray revealed that the heart was within normal limits, there were minimum peri-hilar inflammatory changes but no evidence of parenchymal lung lesions/cavitations.

The child improved with the initial treatment but there was still evidence of persistent lesions on the labial aspect of the gingivae with erythema and hyperplasia over the interdental papillae. He was continued on prednisolone.

He was referred to a consultant paediatrician in a local university hospital to evaluate possible multisystem organ involvement but he was symptom free. After treatment with prednisolone for 3 months the gingival hyperplasia resolved completely. He remained on a maintenance dose of 1 mg of prednisolone for 1 year and did well. He is still under regular review with the paediatrician for monitoring of any possible multi-system involvement.

Oral findings

The aforementioned presenting complaints and subsequent oral findings in relation to Wegener's disease as 'strawberry gums' are very clearly depicted in Fig. 1.

Discussion

Wegener's granulomatois was first described by Friedrich Wegener in 1936[2] and later in 1954 Godman and Churg[3] established the three main clinical criteria of the disease which predominately involved the upper airway and related structures (E), the lungs (L) and the kidneys (K). So the classic form the disease is designated as ELK[4].

The aetiology of the disease is unknown. However, circumstantial evidence supports the theory that it is an autoimmune disease. It is also thought that ANCA play a role. For a disease in which the average untreated survival is less than 1 year, it is very crucial that the oral manifestations, although rare, are recognised before multi-system involvement occurs.

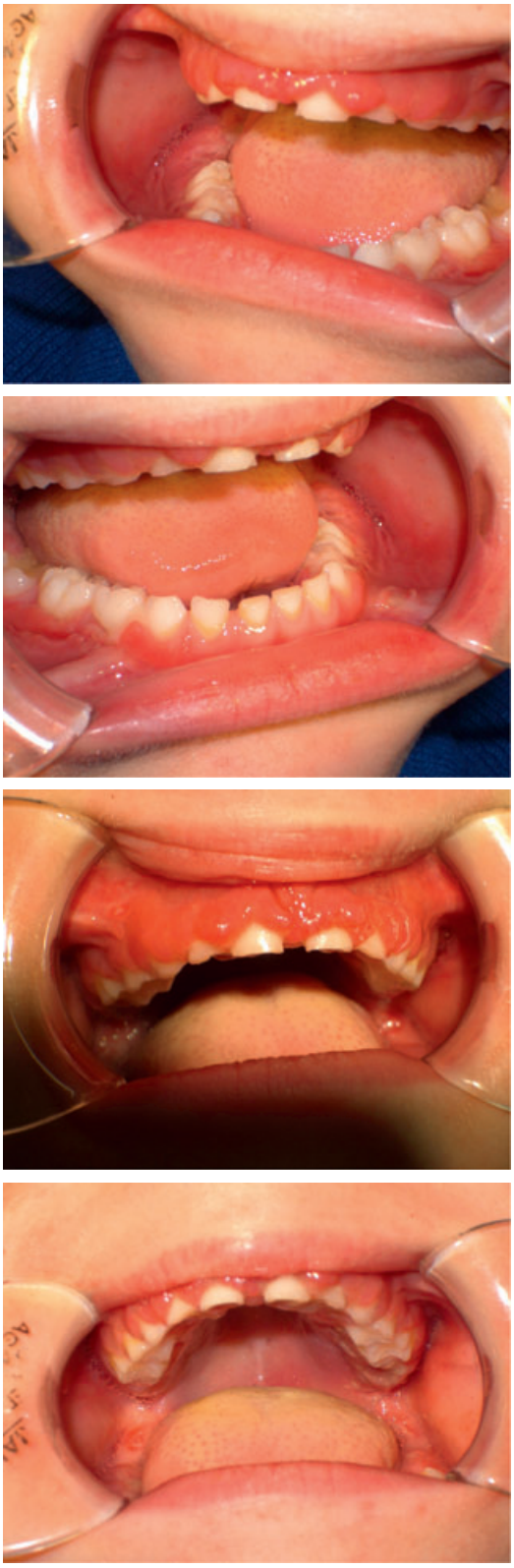

Figure 1 Four cases of strawberry gums.

A typically described triad of full blown WG consists of the following[5]

- necrotising granulomatous inflammation of upper and lower respiratory tracts;
- systemic vasculitis of small arteries and veins;
- focal glomerulonephritis.

However, not all patients show involvement of all three areas and virtually any area can be involved.

Oral or pharyngeal involvement occurs in 6% of the patients but oral lesions as initial signs are rare. The most common oral lesion is friable granular-hyperplastic gingivitis, referred to as 'strawberry gums'. There can be associated alveolar resorbtion and tooth mobility[6]. The other oral findings are persistent mucosal ulcerations usually buccal but may also occur of the palate or pharynx. The disease may remain localised to the mouth for several weeks or months before multi-organ involvement occurs.

Oral histology is important for early diagnosis[7]. The characteristic features are vasculitis and necrotising granulomas.

Gingival enlargement is a direct manifestation of WG and it may be pathogonomic.

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