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Strawberry gingivitis as an initial sign of occult Wegener's granulomatosis - A case report

[1]Dr. Ameena M S, Junior Resident, Dept. of Oral Medicine and Radiology, Government Dental College, Thiruvananthapuram, Kerala, India.

[2]Dr. Sunu Ramachandran, Associate Professor, Dept. of Oral Medicine and Radiology, Government Dental College, Thiruvananthapuram, Kerala, India.

[3]Dr. Girija K L, Associate Professor, Dept. of Oral Medicine and Radiology, Government Dental College, Thiruvananthapuram, Kerala, India.

[4]Dr. Tinky Bose, Professor and HOD, Dept. of Oral Medicine and Radiology, Government Dental College, Calicut, Kerala, India.

[5]Dr. Ramesh S, Assistant Professor, Dept. of Oral Medicine and Radiology, Government Dental College, Thiruvananthapuram, Kerala, India.

[6]Dr. Aashmi Pradeeba Das T H, Junior Resident, Dept. of Medicine and Radiology, Government Dental college, Thiruvananthapuram, Kerala, India.

Corresponding Author: Dr. Ameena M S, Junior Resident, Dept. of Oral Medicine and Radiology, Government Dental College, Thiruvananthapuram, Kerala, India.

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Abstract:

Background: Granulomatosis with polyangiitis (GPA), formerly referred to as Wegener's granulomatosis, is a rare systemic disease of unknown etiology which can affect all areas of the body, including the oral cavity. The typical oral manifestations occur as nonspecific erosive or ulcerative lesions of the oral cavity or appear with hyperplastic gingivitis, so called "strawberry gingivitis".

Case presentation: In this case report, we present a rare case of Wegener's granulomatosis with strawberry gingivitis in a 36-year-old female reported to our OPD complaints of generalized swelling on upper and lower gums since 3 weeks. The patient underwent oral prophylaxis followed by incisional biopsy. We referred the patient to Department of Respiratory Medicine. Based on the histological findings and chest CT a diagnosis of wegenears granulomatosis (GPA) was made. Oral lesions were completely subsided within 1

month on treatment with Prednisolone and Azathioprine. Patient had been followed for 2years without any medication. There was no lesions or symptoms on follow up period.

Conclusion: GPA is a multisystem disorder associated with considerable morbidity and mortality if not treated. The first manifestation of the disease can be seen in the oral cavity. It is important that dentists recognise the oral manifestation in order to improve the prognosis.

Keywords: Granulomatosis with polyangitis, Orofacial mani festation, Straw berry gingivitis, Wegener's granulo matosis.

Introduction

Wegener's granulomatosis is an uncommon multi-organ disease first categorized as a distinct syndrome by Friedreich Wegener in 1936. The hallmarks of this potentially fatal disorder are necrotizing granulomatous inflammation involving the upper and lower respiratory tract, glomerulonephritis, and vasculitis.[1] Wegener's granulomatosis may occur as a limited or generalized disease. The limited form of Wegener's granulomatosis runs an indolent course whereas the disseminated disease has a rapid progressive course leading to life-threatening multi-organ failure. Of the 2 types of Wegener's granulomatosis, patients with the generalized disease are known to have shorter life expectancy than those presenting with the limited disease. Wegener's granulomatosis has an insidious onset and usually develops over a period of time with the mean period from onset of symptoms to diagnosis ranging from 4.7 to 15 months.[2] Without treatment it is invariably fatal and most patients do not survive more than a year after diagnosis. Delay in the diagnosis of Wegener's granulomatosis is attributed mostly to the nonspecific presenting signs and symptoms associated with the early phase of the disease.[3] The most characteristic oral lesion is hyperplastic gingivitis, which is typically red to purple with many petechiae (strawberry gingivitis). These lesions may remain localized in the oral cavity for unusually long periods of time before multi-organ involvement occurs.[4] Therefore, timely recognition of this often-overlooked oral finding can help to establish an early diagnosis of this disease. Management with appropriate therapy produces a good response in most cases, with only occasional relapses.[5]

Case Report

A 36-Year-old female reported to Oral medicine & Radiology department, GDC, TVM with complaints of generalized swelling of upper and lower gums of 3 weeks duration. (Figure 1) The patient had no relevant medical history and no known drug allergy. No significant abnormality detected on reviewing the systems. On intra oral examination there is diffuse, hypertrophic, reddish, friable gingiva with loss of stippling in maxillary (16 to 26) and mandibular (35 to 45) region. (Figure 2 & Figure 3) In maxillary arch palatal gingiva is uninvolved, but in mandibular arch both labial and lingual gingiva is affected. Gingiva is tender on palpation and easily bleed on touch. There was grade II mobility on mandibular anterior teeth. No other lesions were found elsewhere in the oral cavity. Clinical diagnosis of gingival manifestation of granulomatous disease was suspected. Panoramic radiograph revealed generalized horizontal bone loss which was pronounced on mandibular anterior region. (Figure 4) Blood investigation showed elevated neutrophils, elevated ESR & Positive c-ANCA. Mantoux test was negative. Renal Function test results were within normal limits. After oral prophylaxis patient was referred to Department of respiratory medicine for systemic evaluation. A decision was made to perform an Incision biopsy on the gingival growth under local anesthesia. The specimen obtained

was submitted for routine processing followed by staining with H-E, PAS, Gorcott methinamine silver and Ziehl-Nelsen. Microscopic examination revealed ulcerated stratified squamous epithelium and connective tissue stroma. Also showed granulomatous areas with epitheliod cells, intense collections of neurophils, macrophages, plasma cells, localized areas of necrosis & multinucleated gaint cells. (Figure 5) Chest CT axial section showed soft tissue density nodular lesion in the oblique fissure suggestive of granuloma in lung. (Figure 6) MRI and Ultrasonography of liver, gall bladder, pancreas, kidney and urinary bladder appears normal. Based on the histological findings, radiographical findings and the presence of strawberry gingivitis, a diagnosis of Wegner's granulo matosis was made. Patient had been treated with prednisolone 20mg and azathioprine 50mg for 1month. Intra oral examination revealed complete remission of lesions after 1 month. (Figure 7& Figure 8) Patient had been followed for 2 years without any medication. There was no lesions or symptoms on follow up period.

Figure 1: Extra oral photograph

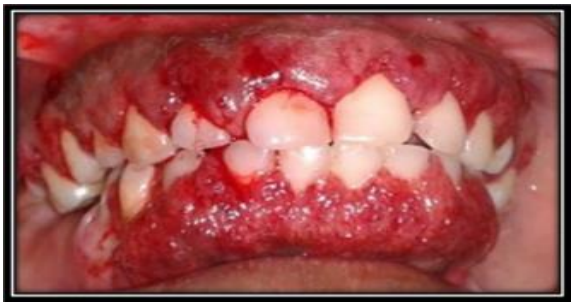

Figure 2: Diffuse, hypertrophic, reddish, friable with loss of stippling in maxillary (16 to 26) gingiva.

Figure 3: Diffuse, hypertrophic, reddish, friable with loss of stippling in mandibular (35 to 45) gingiva.

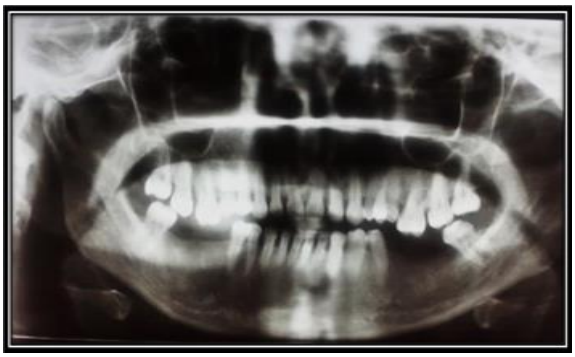

Figure 4: Panoramic radiograph of the patient shows generalized horizontal bone loss with evident bone destruction in the mandibular anterior region.

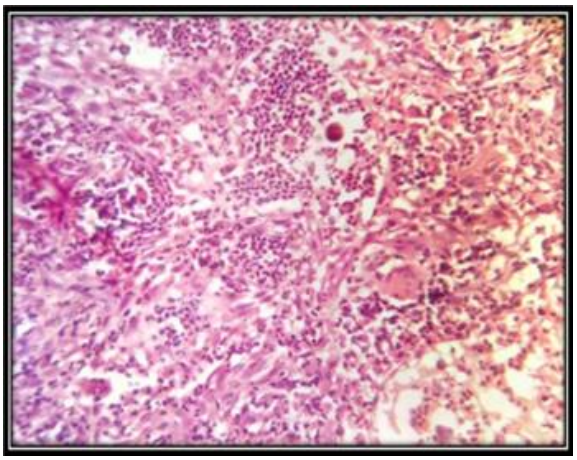

Figure 5: Microscopic examination shows ulcerated stratified squamous epithelium and connective tissue stroma with epitheliod cells, intense collections of neurophils, macroplages, plasma cells, localized areas of necrosis & multinucleated gaint cells.

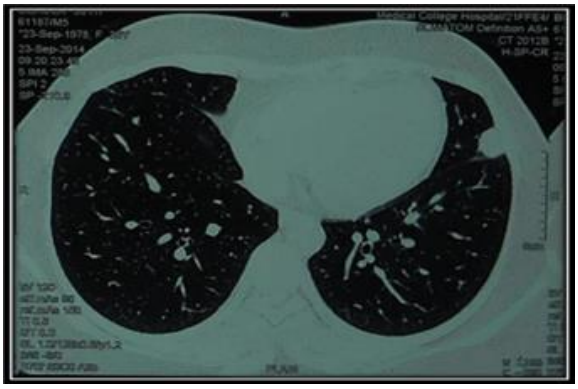

Figure 6: Chest CT axial section shows soft tissue density nodular lesion in the oblique fissure suggestive of granuloma in lung.

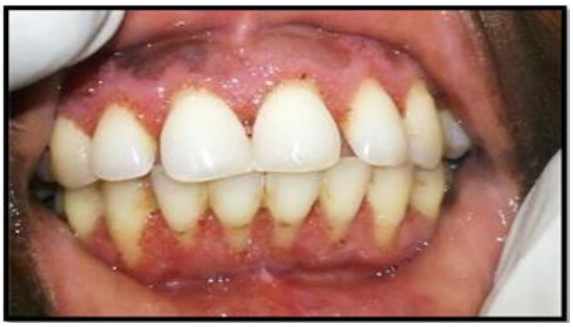

Figure 7: Complete resolution of the maxillary gingival lesions after 1 month post treatment.

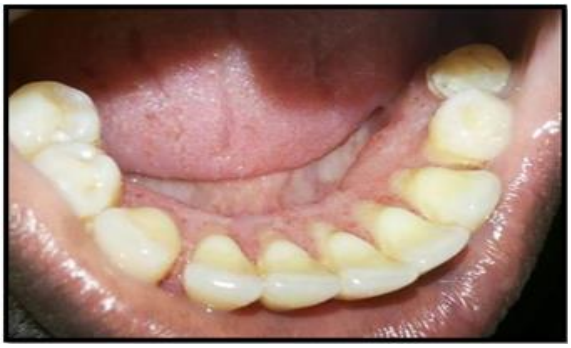

Figure 8: Complete resolution of the mandibular gingival lesions after 1 month post treatment.

Discussion

Wegener's Granulomatosis (WG) is a rare form of systemic vasculitis that can affect the upper respiratory tract, lungs, kidneys and other organs. The head and neck region is usually not spared of the disease either. Only 6% of the cases were presented with oral manifestation, which were in advanced stage of the disease and are rarely an indicator of the disease. Peter McBride, a Scottish otolaryngologist, first described the condition in a paper entitled "Photographs of a case of rapid destruction of the nose and face" in 1897.[6] An earlier name for the disease was pathergic granulomatosis. Although it was Heinz Klinger who first described the disease in two patients who succumbed to widespread sepsis in 1931 in the German literature, the disease is named after Dr. Friedrich Wegener, who described a similar clinical presentation in three patients in 1936. Wegener reported a triad of signs that consisted of necrotising granulomatous inflammation of the upper or lower respiratory tract, systemic necrotising vasculitis of small arteries and veins, and rapid necrotising glomerulonephritis leading to renal failure. The American College of Rheumatology (ACR) recommended that the diagnosis of Wegener's granulo matosis can be made if two of the following criteria are fulfilled:1) ulcerative lesions in oral mucosa or nasal bleeding or inflammation, 2) nodules, fixed infiltrates or cavities in chest radiograph, 3) abnormal urinary sediment and 4) granulo matous inflammation on biopsy.[7] Otorhinolaryn go logical involvement is more common with destruction of nasal septum, resulting in palatal perforation and progressive sensorineural hearing loss. Ophthalmic symptoms may manifest as epiphora due to involvement of the nasolacrimal duct.[8] The oral lesions may manifest either as mucosal ulcer on the tongue, buccal mucosa, gums and palate, or as gingival hyperplasia with classical "strawberry gingivitis. However, Caw son suggested that other lesions may also occur, such as ulceration of the palate by extension from the nose, where destruction of the nasal septum may develop.[1] It may also occur as small ulcer-like aphthae, diffuse ulcerative stomatitis and spontaneous exfoliation of the teeth.

Crohn's disease and sarcoidosis, deep fungal infection (candida, histoplasmosis and Para coccidioidomycosis), tuberculosis, other granulomatous infections like midline lethal granuloma, midline NK/T-cell lymphomas, other anti-neutrophil cytoplasmic antibody (ANCA)-positive vasculitis, drug-induced gingival enlargement and, rarely, cicatricial pemphigoid (CP) or mucous membrane pemphigoid should be included in the differential diagnosis of WG. WG is fatal if left untreated. The mean survival is reported to be 5 years in untreated cases. In the present case, diagnosis of WG was made according to the criteria given by the ACR after careful exclusion of the above-mentioned lesions by appropriate systemic evaluation and lab investigation. Criteria 1 (ulcerative lesions in oral mucosa- strawberry gingivitis) 2 (nodules, fixed infiltrates or cavities in chest radiograph) and 4 (granulomatous inflammation on biopsy) were met. Furthermore, routine blood analysis showed elevated neutophils, elevated ESR & Positive c-ANCA.

Conclusion

Almouhawis et al. stated that WG has a wide range of non-specific characteristics as it can affect multiple systems, making early diagnosis difficult. WG affecting oral mucosa is extremely rare. Identification, early diagnosis and prompt treatment of WG are important in order to get a better prognosis in this potentially lethal disease. Dental surgeons often being the first to examine the oral cavity should be familiar with the classic oral manifestations, diagnostic parameters and treatment modalities of WG. The appearance of strawberry gingiva is unique, distinct and pathognomonic, which makes it easily identifiable by an expert dental practitioner especially oral medicine specialist.

Declaration of patient consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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