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CLINICAL REPORT

A Rare Presentation of Wegener's Granulomatosis in ENT

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Abstract Wegener's granulomatosis or Granulomatosis with polyangiitis is a rare multisystem autoimmune vasculitis disease which may become life threatening due to the various system involvement. Granular hyperplastic gingivitis, the so called "strawberry gingivitis" is a rare manifestation of this disease, yet pathognomic when present. Early diagnosis and treatment is very important.

Keywords Wegener's granulomatosis · Strawberry gingivitis · Vasculitis · C ANCA · Biopsy

Introduction

The uncommon multi-organ disease, Wegener's granulomatosis was first categorized as a distinct syndrome by Friedreich Wegener in 1936. [1,2] This disease, now known as Granulomatosis with polyangiitis is a rare autoimmune disease that can affect people of all ages and men and women are equally affected. It classically involves a triad of upper airway, lungs and renal disease [3]. The hallmarks of this disease are granulomatous inflammation involving the upper and lower respiratory tract, glomerulonephritis, and vasculitis affecting small to medium sized vessels [4,5]. The most common site of involvement at presentation is head and neck in 73–93% of cases [3]. The most common features are rhinitis and sinusitis, including the feeling of blockage due to nasal mucosa edema, purulent or bloody chronic nasal discharge and nasal crusting. The inflammatory process in the facial area can cause skeletal structures destruction with saddle nose and oronasal fistulas formation in severe cases. Oral lesions are seen in only 6–13% of these patients, with first manifestation being limited to gingival inflammation, called the "strawberry gingivitis" [6]. Timely diagnosis of this disease helps in providing appropriate treatment without delay and to reduce the associated morbidity and mortality. We hereby report a case of a 37-year-old female patient who presented with oral lesions. Informed consent was obtained from the patient.

Case Report

A 37 year old female presented with complaints of burning sensation and reddish lesions in mouth since 1 month and ulcer over right side of cheek since 3 weeks. She developed burning sensation in mouth over left upper side while brushing teeth which progressed to involve right upper side of mouth and then the entire mouth in the following days. At the same time, she noticed redness in these sites which bleeds on touch which was also progressive in nature. 1 week following this, she noticed a small nodular swelling over right cheek. Then she consulted nearby hospital and was treated conservatively with vitamin tablets and gel for local application. But in the following days, her symptoms aggravated. Redness progressed to involve entire gums of oral cavity and the nodule over right cheek which burst open with bloody discharge to form an ulcer. As there was no relief of symptoms after consulting multiple local hospitals, patient visited us with the above-mentioned complaints.

The positive examination findings were reddish strawberry coloured granular hyperplasia of upper and lower

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gingiva seen in its entire extent which bleeds on touch and was non-tender. There was also a 2 × 1 cm single ulcer seen over right cheek, oval shape, well defined margines, undermined edge, floor covered with pale granulation tissue and slough. Also, small 0.5 × 0.5 cm ulcer was present over right forehead (Fig. 1).

We admitted the patient, after detailed history and clinical examination, made a provisional diagnosis of Wegeners granulomatosis and performed necessary investigations like routine blood investigations, other blood ivestigations like C ANCA and radiological evaluation, and biopsy to confirm our diagnosis.

The positive investigations were an elevated CRP (58.56 mg/l), positive C ANCA values (> 200RU/ml), CT thorax showed small centrilobular nodules seen in superior segment of right lower lobe and posterior basal segment of left lower lobe and the histopathology was reported as small vessel vasculitis with granulomatous reaction, the diagnosis of Wegener's Granulomatosis can be considered (Fig. 2).

Based on these findings, the patient was treated with tapering doses of steroids, weekly methotrexate 7.5 mg OD and daily folic acid supplementation. We could see improvement in her symptoms in 5 days of starting the treatment (Fig. 3).

Discussion

Granulomatosis with polyangiitis, formerly known as Wegener's granulomatosis is a rare systemic vasculitis associated with the presence of ANCA in serum of patients. Various theories have been postulated regarding the etiopathogenesis of this disease which includes autoimmune theory, the hypersensitivity theory and the theory of infection as a precipitating factor [7]. The autoimmune theory is most accepted and C ANCA is specific for proteinase 3 (PR3), an endogenous peptide.

This disease is characterized by granulomatous inflammation of respiratory tract and necrotizing vasculitis affecting small to medium sized vessels with focal or proliferative glomerulonephritis. The incidence of this disease is estimated to be 10–15 per million per year [3]. Though all ages can be affected, the mean age at diagnosis is 40–50 years and both sexes are equally affected. But in children, female preponderance is observed [3,8].

Strawberry gingivitis, the characteristic sign of GPA, manifests as enlarged, erythematous interdental papillae with red to purple gingival tissue containing petechiae and a granular appearance with yellow punctate foci [9]. In our case, oral complaints and strawberry gingivitis was the first clinical manifestation.

Some of the differential diagnosis for reddish hyperplastic gingival lesions are plaque related inflammatory gingival disease, peripheral giant cell granulomas, haemangiomas, pyogenic granulomas, drug induced gingival hyperplasia secondary to use of dilantin, cyclosporine or nifedipine therapy, Kaposi's sarcoma etc. Thus, proper diagnosis is essential for proper management.

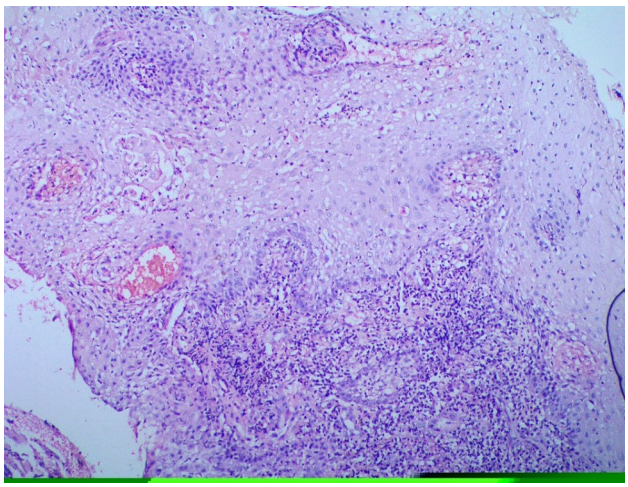

Fig. 2 Histopathology slide showing inflammatory cell infiltrates and vessel wall fibrinoid necrosis

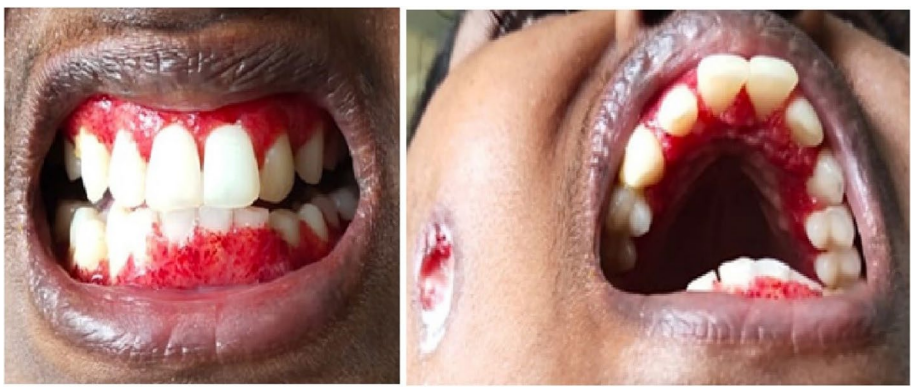

Fig. 1 Oral lesions and ulcer over right side of cheek

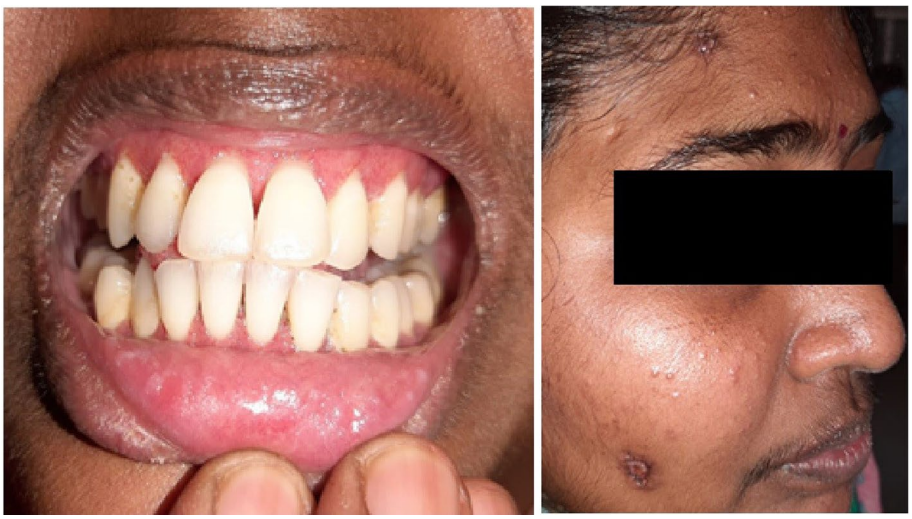

Fig. 3 Almost complete clearance of gingival hyperplasia and healed right cheek and forehead ulcer

As per the American College of Rheumatology (ACR), the diagnosis of Wegener's granulomatosis 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) granulomatous inflammation on biopsy [10,11]. In the case report that we presented, criteria 1,2 and 4 were met. However, the diagnosis is mainly based on biopsy and supportive diagnosis of presence of CANCA in patient's serum.

Histopathologically, GPA disease shows leucocytoclastic vasculitis of small vessels that may demonstrate fibrinotic necrosis in their walls, ill-defined granulomas with multinucleated giant cells, macrophages, lymphocytes and plasma cells, and also extensive areas of necrosis. However, these findings mentioned are more frequently observed in lung biopsies, whereas oral lesions typically reveal intense mixed inflammation with presence of microabscesses, multinucleated giant cells and demonstrates pseudoepitheliomatous hyperplasia. [12,13] For our patient, we had taken biopsy from the oral lesion, both from upper and lower gingiva and they demonstrated fibrinoid necrosis of vessel wall and granulomas with inflammatory infiltrates.

The course of GPA is usually chronic with rapid progression. As it can be life-threatening leading to organ failure, early diagnosis and proper treatment is crucial for the patient's better prognosis. Mild disease with no evidence of active disease (i.e.; normal serum creatinine and no red cell casts or proteinuria) and no organ threatening manifestations are treated with glucocorticoids in combination with either rituximab or methotrexate. Moderate to severe disease is treated with glucocorticoids in combination with either cyclophosphamide or rituximab [3]. Our patient was treated with tapering doses of steroids and methotrexate and we were able to see significant improvement in symptoms and clinical manifestation in few days of starting the treatment. The patient is on regular follow up, with no recurrence till date.

Conclusion

Wegener's granulomatosis is a rare autoimmune disease with multisystem involvement. Negligence is not advocated as it can be life threatening. Long term treatment is required. Recurrences are common and hence, long term follow up is advised.

Author Contributiion Raghavendra Prasad K.U.–concept, design. Fida Harish A.T.–literature search, data acquisition, manuscript preparation and editing. The manuscript has been read and approved by all the authors, the requirements for authorship have been met, and each author believes that the manuscript represents honest work.

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Declarations

Conflict of interest None.

Ethical Approval Institutional research committee and Institutional ethical committee approval obtained.

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