

## Case Report

# Wegener's Granulomatosis presenting as a small soft palate mass

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### Abstract:

*Wegener's granulomatosis (WG) is a form of vasculitis involves small and medium-sized blood vessels. It commonly involves the upper and lower respiratory tract. However, multisystemic involvement and involvement of kidneys are possible and may lead to a life threatening condition. Therefore early diagnosis is important. Timely beginning of clinical management may considerably influence the further course of disease. Although, it may present as oro-pharyngeal mass surgical procedure is not indicated method of treatment of Wegener's granulomatosis, because it can increase the pathologic process. Here we report a case of Wegener's granulomatosis (WG) presenting as a tissue growth over the soft palate.*

**Key words:** ANCA, Vasculitis, Wegener's granulomatosis

### Introduction:

Wagener's granulomatosis (WG) is a small—and middle--vessels vasculitis. The pathomorphological diagnostic criteria is known as Wagener's triad: 1) necrotizing granlomatous inflammation of upper and/or lower respiratory tract, 2) systemic or focal necrotizing vasculitis involving arteries and vein, and 3) focal segmental necrotizing crescentic glomerulonephritis<sup>1,2</sup>. WG is kind of vasculitis where multisystemic involvement and involvement of kidneys are possible and may lead to a life threatening condition. We describe a case of a 35-year-old man with symptoms of a mass in the soft palate who subsequently diagnosed Wagener's granulomatosis on the basis of histopathology.

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### Case report:

A 35-year-old man presented for consultation at Green Life Medical College Hospital, Dhaka, with pain in the throat and difficulty in swallowing for 15 days. He had no fever. He did not give any history of epistaxis, haemoptysis, haematemesis or weight loss. There was no history of a skin rash. He did not give any significant recent illness other than present condition. Oral examination revealed a 2-3 cm diameter patchy tissue growth on the soft palate with ulceration, oval in shape, floor is inflamed with whitish slough and margin is not well defined. The gums and remainder of the oral cavity were normal in appearance.



**Figure 1:** Showing lesion over the soft palate.

Investigation include ESR-40mm 1<sup>st</sup> hour. Chest x-ray normal, creatinine 0.9 mg/dl, c-ANCA (ELISA Assay) was borderline positive (20 U/ml). P-ANCA was negative. Random blood glucose level was 6.1 mmol/L. Urine R/E was normal.

Small piece of tissue has been excised and submitted for histopathological examination. Microscopy showed active vasculitis, necrotising granulomatous inflammation consistent with Wegener's granulomatosis.

The disease is clinically localized in oral cavity. Therefore, it is a limited form of WG rather than severe one. The patient was discharged with oral prednisolone 40 mg advised to come after 15 days for follow-up. Patient may need disease modifying drugs like Cyclophosphamide or Azathioprine on the basis of disease progression and systemic involvement in subsequent follow up.

#### **Discussion:**

WG was first described by Klinger in 1933, followed by other investigators, including Rossle in 1933, Wegener in 1936 and 1939, and Ringertz in 1947<sup>3,4</sup>.

WG is classified as ANCA positive vasculitis, mostly localized on the small and medium-sized blood vessels. It mostly affects the upper and lower respiratory airways and kidneys<sup>5</sup>. According to literature data, the lungs are affected in 90 percents of patients<sup>6</sup>. Typical radiological presentations of the lung involvement are multiple, bilateral, nodular infiltrations, with or without cavities. According to some data, in 20-50% of patients it is manifested with pleural effusion<sup>6</sup>. Atypical presentations are interstitial lung disease, hilar mass or pneumothorax<sup>7,8</sup>. In 1990, the American College of Rheumatology (ACR) established the criteria for the classification of WG<sup>9</sup>: 1) nasal or oral inflammation, 2) radiologically demonstrated pulmonary infiltrates, 3) abnormal urinary sediment (red

cell cast, haematuria), 4) granulomatous inflammation on biopsy. Patient shall be said to have Wegener's granulomatosis if at least 2 of these 4 criteria are present. The presence of autoantibodies to proteinase 3/ cANCA is not required for diagnosis of WG, by either ACR or Chaper Hill consensus Conference (CHCC) definition<sup>9</sup>. Occasionally, patients with infection, inflammatory bowel disease, rheumatic disease, neoplasm develop ANCA<sup>10</sup>.

Because oral lesions may be the presenting manifestation of WG, dentists, oral and head and neck surgeons, and internal medicine specialists should be familiar with the clinical features. A complete evaluation, with biopsies, medical management, close monitoring are necessary to minimize morbidity and prevent mortality. Several oral manifestations of WG have been described, including a characteristic hyperplastic gingivitis termed 'strawberry gums', clinically apparent facial swelling and oral ulceration<sup>11,12</sup>.

Our case showed only patchy tissue growth in palate, raising a differential diagnosis including infectious, reactive and neoplastic disease. An experience pathologist can rule out bacterial or deep fungal infection while histological examination of lesional tissue. Similarly, neoplastic disease can be excluded by means of biopsy. Biopsy specimens need to be adequately deep to establish a definite diagnosis of WG. According to some author cANCA estimation remains the definitive diagnostic test for WG. But both the immunofluorescent and ELISA forms of analysis tests for cANCA may be negative in a significant proportion of cases. Therefore, care should be exercised in the interpretation of results. In our case cANCA was 20 U/ml. As our case was a limited form of Wegener's Granulomatosis, he was given only oral prednisolone and on follow-up to see the disease process<sup>13, 14, 15</sup>.

**Conclusion:**

Disease of the upper airway is a common presenting manifestation of Wegener's granulomatosis. Nasal or oral ulceration, epistaxis, sinusitis, or otitis is found in more than 90% of cases. Therefore, Wegener's granulomatosis remains a challenging clinical problem. Early diagnosis and the timely beginning of clinical management may considerably influence the further course of the disease. Surgical procedure is not indicated method of treatment of Wegener's granulomatosis, because it can increase the pathologic process. So, clinician should be aware of early diagnosis and management of WG to prevent systemic involvement.

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