CASE REPORT

Wegener's Granulomatosis with a Difference

Nithin Thomas[1], Vishwambar Vallabaneni[2], Jereen Varghese[3], Ragulan R[4], Sreekaanth Sundarraj[5]

Abstract

A 60-year-old male presented with complaints of hemoptysis, cough, chest pain, nasal crusting, and hematuria. He had history of diabetes for the last 20 years on orally administered antihyperglycemic agents (OHAs). He was a smoker for the past 30 years. On examination, patient had strawberry gingival hyperplasia. Auscultation revealed bilateral wheeze on all areas of chest. High-resolution computed tomography (HRCT) revealed ill-defined multifocal patchy areas of airspace consolidation in both lungs involving the upper and lower lobes. Rheumatoid arthritis (RA) factor, antinuclear antibody (ANA) enzyme-linked immunoassay (ELISA), anti-cyclic citrullinated peptide (CCP) antibodies ELISA, deoxyribonucleic acid (DNA) (double strand) antibody were negative, angiotensin-converting enzyme (ACE) was within normal limits while cytoplasmic anti-neutrophil cytoplasmic antibody (C-ANCA) was positive and perinuclear anti-neutrophil cytoplasmic antibody (P-ANCA) was negative. A diagnosis of granulomatosis with polyangiitis was made in accordance with the American College of Rheumatology Criteria for Wegener's granulomatosis. Strawberry gingival hyperplasia is a rare but pathognomonic manifestation of granulomatosis with polyangiitis (GPA); hence I present this case report.

Keywords: Cytoplasmic anti-neutrophil cytoplasmic antibody, Granulomatosis with polyangiitis, Vasculitis, Wegener's granulomatosis.

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Introduction

GPA stands for granulomatosis with polyangiitis, a rare multisystem autoimmune illness characterized by necrotizing granulomatous vasculitis. The traditional triad of GPA includes upper respiratory involvement (chronic sinus infections, nasal mucosal lesions, otitis, subglottic stenosis, and skeletal abnormalities), lower airway tract involvement (chest discomfort, cough, and hemoptysis), and glomerulonephritis.[1] The disease begins as localized granulomatosis of the respiratory tract, then progresses to a vasculitis that primarily affects small- and medium-sized arteries.[2] Presentation of the patient depends on the organ which is affected. When patients appear with chronic nasal obstruction, they may be misinterpreted as having chronic sinusitis, while others may come with severe respiratory or renal failure. The most common respiratory complaints are cough with or without hemoptysis, chest discomfort, fever, and dyspnea.[2]

Case Description

Presenting a case of GPA in a 63-year-old male with chief complaints of hemoptysis, cough, chest pain, nasal crusting, and hematuria; H/o diabetes for 20 years on OHAs; no previous history of pulmonary tuberculosis/bronchial asthma; and smoker for the past 30 years. On examination, patient had strawberry gingival hyperplasia. Auscultation revealed bilateral wheeze on all areas of chest. Chest X-ray was normal (Fig. 1).

High-resolution computed tomography (HRCT) revealed ill-defined multifocal patchy areas of airspace consolidation in both lungs involving the upper and lower lobes. Air bronchogram and multiple small cystic/pseudocavitating foci were seen within the airspace opacities. The lesion shows central more than peripheral with peribronchovascular distribution (Fig. 2).

Bronchoscopy revealed no abnormality; bronchoalveolar lavage (BAL) analysis was normal. Rheumatoid arthritis factor (RA factor), antinuclear antibody (ANA) enzyme-linked immunoassay (ELISA), anti-cyclic citrullinated peptide (CCP) antibodies ELISA, deoxyribonucleic acid (DNA) (double strand) antibody were negative, angiotensin-converting enzyme (ACE) was in normal limits while cytoplasmic anti-neutrophil cytoplasmic antibody (C-ANCA) was positive, and perinuclear anti-neutrophil cytoplasmic antibody

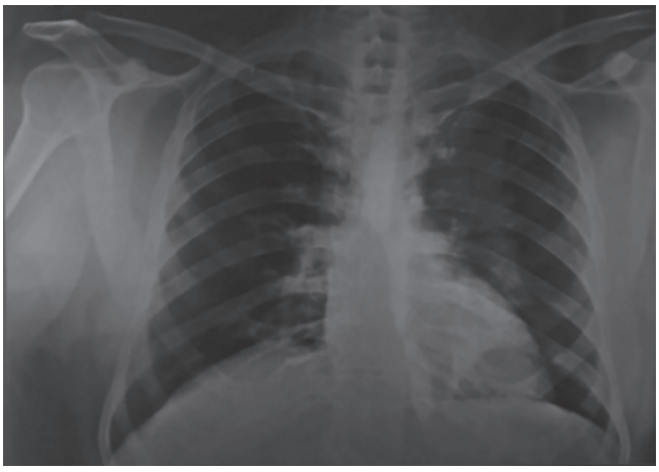

Fig. 1: Chest X-ray

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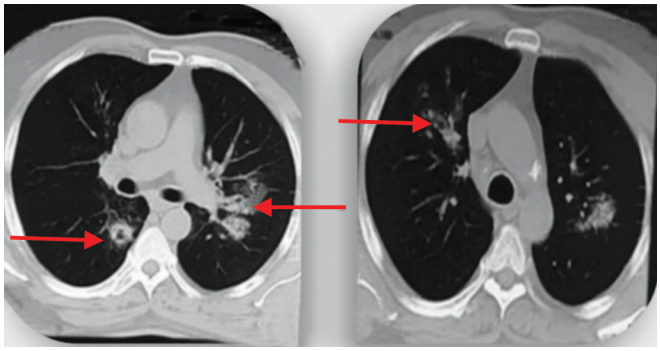

Fig. 2: HRCT

(P-ANCA) was negative. C-reactive protein (CRP) was elevated and urine routine revealed 8–10 RBCs/HPF. A diagnosis of GPA was made in accordance with the American College of Rheumatology Criteria for Wegener's granulomatosis.[3] He was advised to start on rituximab and corticosteroids.

Discussion

Necrotizing granulomatous inflammation as well as pauci-immune vasculitis of medium-sized vessels are features of GPA. Cytoplasmic components of neutrophils (C-ANCA) are circulating antibodies that have been detected in >90% of patients.[4] GPA is caused by anti-neutrophil cytoplasmic autoantibodies specific for proteinase 3 (PR3-ANCA).[5] Computed tomography (CT) imaging can reveal nodules that cavitate, thin- or thick-walled cavities, airway stenosis, ground-glass opacities secondary to pulmonary bleeding, ground-glass halo ("CT halo" sign) or reverse halo ("atoll" sign), and ulcerations.[6] Even when the radiograph appears normal, CT imaging can detect the disease. In cases, when opacities have previously been discovered in bilateral lung areas on a standard chest radiograph, CT scanning can be used to characterize and distribute them.[7] Wegener's granulomatosis with a "difference"—strawberry gingival hyperplasia even though is rare, it is pathognomonic manifestation of GPA.

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References

1. Frankel SK, Cosgrove GP, Fischer A, Meehan RT, Brown KK. Update in the diagnosis and management of pulmonary vasculitis. Chest 2006;129(2):452–465. DOI: 10.1378/chest.129.2.452.
2. Martinez F, Chung JH, Digumarthy SR, Kanne JP, Abbott GF, Shepard JA, et al. Common and uncommon manifestations of Wegener granulomatosis at chest CT: radiologic-pathologic correlation. Radiographics 2012;32(1):51–69. DOI: 10.1148/rg.321115060.
3. Leavitt RY, Fauci AS, Bloch DA, Michel BA, Hunder GG, Arend WP, et al. The American College of Rheumatology 1990. Criteria for Wegener's granulomatoses. Arthritis Rheum 1990;33(8):1101–1107. DOI: 10.1002/art.1780330807.
4. Lynch JP III, Leatherman JW. Alveolar hemorrhage syndrome. In: Fishman's pulmonary diseases and disorders. 4th ed. vol 1. 2008. p. 77;1289.
5. Jennette JC. Nomenclature and classification of vasculitis: lessons learned from granulomatosis with polyangiitis (Wegener's granulomatosis). Clin Exp Immunol 2011;164(Suppl. 1):7–10. DOI: 10.1111/j.1365-2249.2011.04357.x.
6. Ananthakrishnan L, Sharma N, Kane JP. Wegener's granulomatosis in the chest: high resolution CT findings. Am J Roentgenol 2009;192(3):676–682. DOI: 10.2214/AJR.08.1837.
7. Varghese J, Krishnapriya R, Srinivasan R. Granulomatosis with Polyangiitis-A case report. IAIM 2016;3(6):207–210.