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# Atypical radiological findings of Wegener's granulomatosis

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**Abstract**
Wegener's Granulomatosis is a rare and invariably fatal form of systemic vasculitis, but early diagnosis and management have significant positive impact on future outcome and prognosis. Wegener's granulomatosis (WG) is characterized by necrotizing granulomatous lesions of the respiratory tract lower and upper, along with necrotizing glomerulonephritis and widespread disseminated vasculitis. In India, two forms of Wegener's granulomatosis have been increasingly reported namely a limited WG (LWG) involving only upper and lower respiratory tract involvement; and the classical WG (CWG) with both respiratory and renal involvement. Study of this case emphasizes the need for careful consideration and systematic analysis of patient's along with diagnostic investigations.

**Keywords:** Wegener's granulomatosis, Atypical radiological findings

## Introduction
Wegener's granulomatosis is an autoimmune small vessel necrotising vasculitis associated with both granulomatosis and polyangiitis. Wegener's granulomatosis (WG) is characterized by necrotizing granulomatous lesions of the respiratory tract lower and upper, along with necrotizing glomerulonephritis and widespread disseminated vasculitis. The form of WG is designated ''limited'' when only one or two organ systems are involved without renal involvement [1].

Scleritis mostly occurs as a chronic inflammatory process, in some instances although it may be caused directly by an infective agent, in vast majority of cases scleritis presents as an immune-mediated disorder [2-4]. Scleritis has been reported to occur in 16–38% of patients with WG [5]. PUK is typically associated with an underlying systemic disease, most commonly rheumatoid arthritis followed by WG [6].

Comparison of the ACR and EULAR/PRES classification criteria for WG*

1990 ACR criteria A patient is said to have WG when 2 of the following 4 criteria are present: Nasal or oral inflammation Abnormal chest radiograph Abnormal urinary sediment (microhematuria [5 red blood cells per high-power field] or red blood cell casts in urine sediment) Granulomatous inflammation on biopsy EULAR/PRES criteria A patient is said to have WG when 3 of the following 6 criteria are present:

Nasal or sinus inflammation Abnormal chest radiographer chest CT scan Abnormal urinalysis (hematuria and/or significant proteinuria) Granulomatous inflammation on biopsy/necrotizing pauci-immune GN Subglottic, tracheal, or endobronchial stenosis

Anti-PR3 ANCA or cANCA staining Differences between American College of Rheumatology (ACR) and European League Against Rheumatism/Pediatric Rheumatology European Society (EULAR/PRES) classification systems and for individual criteria.

Thus one should suspect Wegener`s granulomatosis even in absence of typical radiological features and additional diagnostic investigations such as positive immunofluorescence should be recognized as reliable and valuable diagnostic tools in the absence of histopathology and constitutional symptoms for the diagnosis and prognosis of Wegener's Granulomatosis. Unusual features and literature on this entity have been discussed.

## Discussion
In India, two forms of wegener's granulomatosis have been increasingly reported namely a limited WG (LWG) involving only upper and lower respiratory tract involvement; and the classical WG (CWG) with both respiratory and renal involvement [7].

WG is a part of a larger group of systemic necrotising vasculopathies which include Churg-Strauss syndrome and microscopic polyangiitis, all of which are believed to be caused by autoimmune attack of an abnormal circulating antibody (ANCA) against medium and small sized blood vessels.[8] In particular, ANCA against Proteinase-3 (PR3-ANCA) detected in blood is highly specific for WG [9].

Multinodular disease of the lung is a common characteristic of metastatic lung disease, associated in particular with renal, breast, thyroid and gastrointestinal tract malignancies. The characteristic radiographic finding of "cannon ball" is multiple, solid, well-circumscribed parenchymal masses of variable sizes [10-12] Numerous, nonmalignant diseases may also present with a similar radiological picture such as cryptogenic organizing pneumonia (COP), tuberculosis, aspergillosis and Wegener's granulomatosis [13-15]

CT scan suggestive of round nodule with irregular margin with cystic spaces and mid-peripheral location, typical features suggesting wegener's granulomatosis.

Its close differentials were

**Metastasis:** Typically metastasis appears of soft tissue attenuation, well circumscribed round lesions and more often in periphery of the lung.

A prominent pulmonary vessel has frequently been noted heading into a metastasis.

1. Tuberculosis: Tree-in-bud sign or pattern describe the CT appearance of multiple area of cetrilobular nodules with a branching pattern.
2. Sarcoidosis: Mediastinal lymph node enlargement and bilateral pulmonary opacities, small nodules mostly along the bronchovascular bundles, gives bronchi and vessels a beaded appearance

Cannon ball appearance in radiology always demands thorough diagnostic workup including searching of the primary. The multiple cannon ball lesions without any established primary site, though quite rare, present a diagnostic challenge as not only the other entities are rare but also the list is unlimited. WG is an elusive disease and has non-specific features particularly in the initial stages and a strong index of suspicion is necessary for early diagnosis which is imperative for institution of early immunosuppressive therapy. In case of atypical presentations, the diagnosis in these patients poses a great clinical problem, bearing in mind a small number of positive pathohistological findings, as well as the possibility of positive ANCA antibodies in some other diseases.

In the absence of early immunosuppressive therapy, the disease spreads rapidly and is usually fatal. It is vital not to miss the diagnosis early because early initiation of immunosuppressive therapy with steroids and cyclophosphamide can lead to overall 5 year-survival rates in excess of 80% [16].

## Conclusions

Wegener's Granulomatosis is a rare and invariably fatal form of systemic vasculitis, but early diagnosis and management have significant positive impact on future outcome and prognosis.

Study of this case emphasizes the need for careful consideration and systematic analysis of patient's along with diagnostic investigations.

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