

# Jacobs Journal of Clinical Case Reports

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Case Report

## Pulmonary Mass as an Unusual Radiologic Presentation of Granulomatosis with Polyangiitis: A Case Report

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Received: 06-15-2015

Accepted: 07-08-2015

Published: 08-11-2015

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### Abstract

A previously healthy 40 year old Mexican mestizo male, was seen at another hospital two years before because of nasal congestion, diplopia, loss of peripheral vision, nocturnal diaphoresis, polyarthralgias and unintentional weight loss. He received oral steroids for 6 months achieving complete resolution of symptoms. He was referred because symptoms recurred a nasal septal perforation was diagnosed as well as bilateral vocal cord paralysis and subglottic stenosis. He went on to develop fever, non-productive cough, pleuritic chest pain, arthralgias, an erythematous rash that involved both legs and diminished visual acuity in the left eye. Initial chest X ray showed a well- defined pulmonary mass in the right upper lobe, so he was admitted because of a probable pulmonary neoplasm. Skin biopsy showed a leucocytoclastic vasculitis and nerve biopsy was consistent with axonal degeneration. Anti-PR3 levels were 24.2 and anti-MPO were non reactive. Initial punch lung biopsy demonstrated a poorly differentiated neoplasm which was not confirmed and an open lung biopsy was performed, it demonstrated non caseificant granulomas. Wegener's granulomatosis diagnosis was made. Induction remission treatment was begun and systemic symptoms disappeared progressively.

**Keywords:** Granulomatosis with Polyangiitis; Radiologic Features

### Abbreviations:

BVAS: Birmingham Vasculitis Activity Score;

ENT: ears, nose, throat;

MPO: myeloperoxidase

## Introduction

According to the 2012 revised criteria Granulomatosis with polyangiitis (GPA), previously known as Wegener Granulomatosis (WG) is a necrotizing granulomatous inflammation of the upper and lower respiratory tracts, with necrotizing of small and medium size vessels (polyangiitis). Necrotizing glomerulonephritis is common but is not essential for the classification. The most common lower respiratory tract symptoms in GPA are cough, hemoptysis (due to alveolar hemorrhage and/or tracheobronchial disease), dyspnea, and pleuritic pain. The severity of symptoms and signs varies considerably from asymptomatic (one-third of patients) to acute and fulminant alveolar hemorrhage with respiratory failure. The specific clinical manifestations vary depending on whether the patient has tracheobronchial disease, lung parenchymal nodules, interstitial lung disease, or alveolar hemorrhage. Common CT findings in GPA include multiple pulmonary nodules (generally less than 10 in number) and ground glass or consolidative opacities that are patchy or diffuse (due to diffuse pulmonary hemorrhage or active vasculitis). Nodules range in size from a few millimeters to 10 cm, approximately 30 to 50 percent of nodules are cavitory and 17 percent are diffuse opacities. Less common findings include a reticular pattern, interlobular septal thickening, honeycombing (in a minority of patients), bronchiectasis, hilar adenopathy, and pleural effusions.

## Case Presentation

A previously healthy 40 year old Mexican mestizo male was admitted to the hospital because of fever, non-productive cough, chest pain, arthralgia, an erythematous rash that involved both legs and diminished visual acuity in the left eye.

Ten years earlier, he had self-remitting diplopia and loss of peripheral vision on the left eye. Two years before admission, he developed intermittent nocturnal diaphoresis and arthralgia that started in knees and ankles and later spread to shoulders and hands. He also lost 22 pounds, but denied fever, cough or hemoptysis. As the symptoms progressed, the patient could barely walk and later on he developed chest pain. He was admitted and treated for pneumonia; during hospitalization he developed bilateral red eyes with photophobia, diminishing visual acuity and epiphora as well as hypoacusia and serous otorrhea in the left ear. He was discharged with a prescription for oral steroids for 6 months achieving complete resolution of symptoms, after that he stopped treatment and remained asymptomatic for one year.

Three months before being admitted, he presented dysphonia, with non-bloody nasal discharge and nose pain. He was seen at an ambulatory setting by an otolaryngology consultant that found a 25 x25 mm anterior nasal septal perforation as well as a bilateral vocal cord paralysis and a 30% subglottic stenosis. Two days prior to the admission, he developed fever, cough,

pleuritic chest pain, arthralgia, an erythematous rash that involved both legs and diminished visual acuity in the left eye.

The temperature was 38.7 C, the pulse was 75 beats per minute, and the respiratory rate was 25 breaths per minute. The blood pressure was 120/80 mmHg and the oxygen saturation 89% while the patient was breathing room air. He was not in respiratory distress, his speech was fluent, but dysphonic; there was a 0.3 in diameter painful ulcer in the left lateral border of the tongue, with clean base and an erythematous throat; no lymph node enlargement, lung consolidation syndrome in right upper lobe, a normal abdomen; and a disseminated rash in both legs characterized by erythematous lesions, well circumscribed, not blanchable with pressure (Figure 1). He had temporal hemianopsia by campimetry in the left eye and hyperesthesia in both anterior peroneal nerve territories. Laboratory studies results are in table 1.



**Figure 1.** Dermic rash. Erythematous rounded lesions, that didn't disappeared with vitropression maneuver.

Chest X ray showed a well- defined pulmonary mass in the right upper lobe, so he was admitted because of a probable pulmonary neoplasm. A chest CT scan and MRI both showed a pulmonary mass (Figure 2) therefore and a punch biopsy was performed.

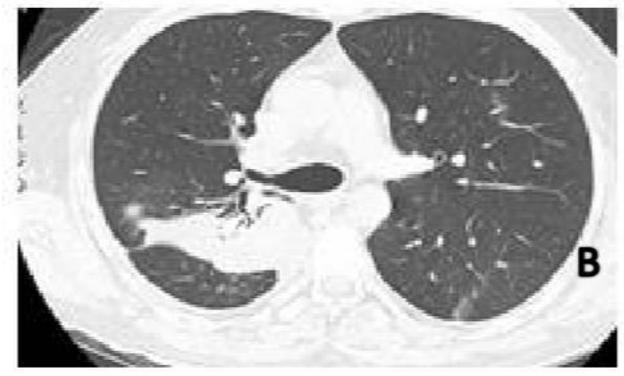
The ophthalmology consultant confirmed the temporal hemianopsia in the left eye, and diagnosed uveitis and episcleritis. Because of hyperplastic gingivitis and pain, a CT scan was ordered evidencing prominent lithic activity and bone destruction. Anti proteinase 3 levels (PR3-ANCA) were 24.2 (negative <5) and anti myeloperoxidase (MPO-ANCA) were non reactive. A skin and nerve biopsy was performed. Skin biopsy showed a leucocytoclastic vasculitis and nerve biopsy was consistent with axonal degeneration. Closed lung biopsy demonstrated a poorly differentiated neoplasm which was not confirmed by

the oncologic consultant that performed an open lung biopsy. Tissue analysis demonstrated non-caseous granulomas. Granulomatosis with polyangiitis was diagnosed was made, the Birmingham vasculitis activity score (BVAS) at diagnosis was 42. Induction remission treatment with high dose methylprednisolone and pulse with Cyclophosphamide/mesna was started.

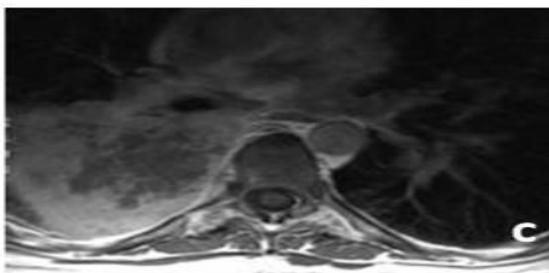
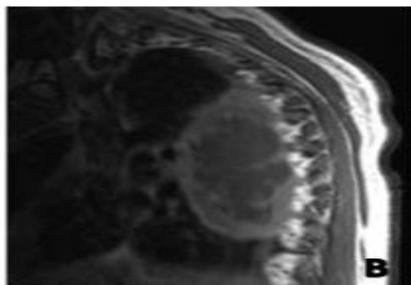
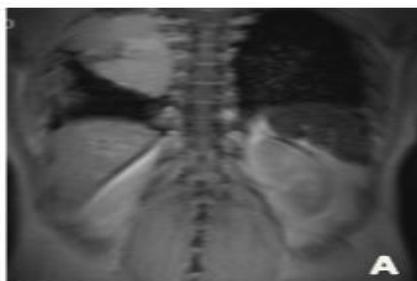
4 months after the induction remission was started, BVAS was 13, and a CT scan demonstrated that the pulmonary mass had reduced its size (Figure 3). Systemic symptoms disappeared progressively. The patient completed induction treatment and is still in consolidation phase, without disease activity.

Parameter		Normal ranges	Parameter		Normal ranges
WBC	12.2 x 10 <sup>3</sup> /μL	4.6-10.2 x 10 <sup>3</sup> /μL	Tryglicerides	143mg/dL	<150 mg/dL
Hb	12 g/dL	13.0-18 g/dL	Total Proteins	8.7 g/L	6.6-8.7 g/L
Plat	448 x 10 <sup>3</sup> /μL	150-400 x 10 <sup>3</sup> /μL	Alb	4.22 g/dL	3.4-4.8 g/dL
Glucose	109 mg/dL	65-110 mg/dL	AST	34 U/Lt	2-38 U/Lt
Urea	22 mg/dL	10-50 mg/dL	ALT	51 U/Lt	2-41 U/Lt
Cr	0.83 mg/dL	0.4-1.2 mg/dL	GGT	20 U/Lt	11-33 U/Lt
Cholesterol	143 mg/dL		DHL	366 U/Lt	240-480 U/Lt
Na	137 mEq/L	136-145 mEq/L	K	4.66 mEq/L	3.5-5 mEq/Lt
Urinary exam	normal				

**Table 1.** Laboratory studies results at admission.



**Figure 3.** Follow up chest computed tomography. Coronal (A) and axial (B) high resolution chest tomography image showing significant reduction of the pulmonary mass 4 months after the induction remission was started.



**Figure 2.** Magnetic resonance imaging (MRI) findings. Coronal (A), sagittal (B) and axial (C) non-contrast-enhanced T1 imaging showing a pulmonary mass in the right upper lobe.

**Conclusion**

Granulomatosis with polyangiitis is a rare systemic disorder, and because it involves multiple organs and may present with unusual radiographic features, affected patients may seek help from physicians in many specialties. Suspicion based in clinical features, early recognition, and prompt treatment can improve clinical outcomes in these patients.

**Discussion**

Granulomatosis with polyangiitis, an uncommon autoimmune disease, is a small-vessel vasculitis. It has systemic involvement but it mainly affects the respiratory tract and kidneys

[1]. Approximately 10-55% of the patients have upper airways involvement [2], and up to 85% develop pulmonary manifestations<sup>1</sup>.

The usual radiographic findings in the lower respiratory tract are lung nodules, interstitial pattern or consolidation, pleural disease and mediastinal lymphadenopathy [3]. The most frequent initial presentation reported in a case series was of multiple cavitory nodules [4], but up to 25% can present with solitary nodules or masses [5]. Although, atypical radiologic features such as a large cavitating lesion has also been reported [6].

In this case, the size of the pulmonary mass and the overwhelming wasting syndrome initially oriented the diagnosis towards a neoplasm. The lack of renal symptoms made the diagnosis even harder. Nonetheless, the recurrent episodes of upper respiratory tract symptoms, the rash and the nerve involvement suggested vasculitis as a differential diagnosis.

Nowadays, granulomatosis with polyangiitis with atypical presentation is still a diagnostic challenge. A high index of suspicion, altogether with signs, symptoms, laboratory results, image and pathological features are needed for a prompt diagnosis.

### Consent

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

### Competing interests

The authors declare that they have no competing interests.

### Authors' contributions

RC, ER and TG wrote this manuscript. GF is the corresponding author, and supervised the manuscript. GF participated in the patients' therapy and helped to draft the manuscript. JMG proofread the final manuscript. All authors read and approved the final manuscript.

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