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# Increased 18F-FDG-PET Uptake in Granulomatosis with Polyangiitis: Case Report and Review of the Literature

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## **Acknowledgements**

N/A

## CASE REPORT

# Increased 18F-FDG-PET Uptake in Granulomatosis with Polyangiitis: Case Report and Review of the Literature

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**G**ranulomatosis with polyangiitis (GPA) is the most common of the antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitides. It frequently involves the kidneys and the upper and lower respiratory tracts.<sup>1</sup> Up to 22% of patients have disease limited to the respiratory tract,<sup>2–4</sup> which presents as diffuse alveolar hemorrhage (DAH) in 5–45% of cases.<sup>1, 5</sup> Tracheobronchial/endobronchial disease is not uncommon, occurring in 10–15% of cases.<sup>2</sup> Hilar adenopathy and mediastinal masses are rare in patients with GPA, as a retrospective study of 302 patients reported a frequency of 2%.<sup>6</sup> We describe an unusual case of GPA presenting with a right lower lobe (RLL) mass and hilar/mediastinal adenopathy.

## CASE PRESENTATION

A 70-year-old woman with a smoking history presented with right shoulder pain and pleuritic chest pain. She denied cough, dyspnea, hemoptysis, and constitutional symptoms. Vital signs and physical exam were unremarkable. Laboratory data (including urinalysis and renal function tests) were normal. She reported recent air travel, raising concern for pulmonary embolism.

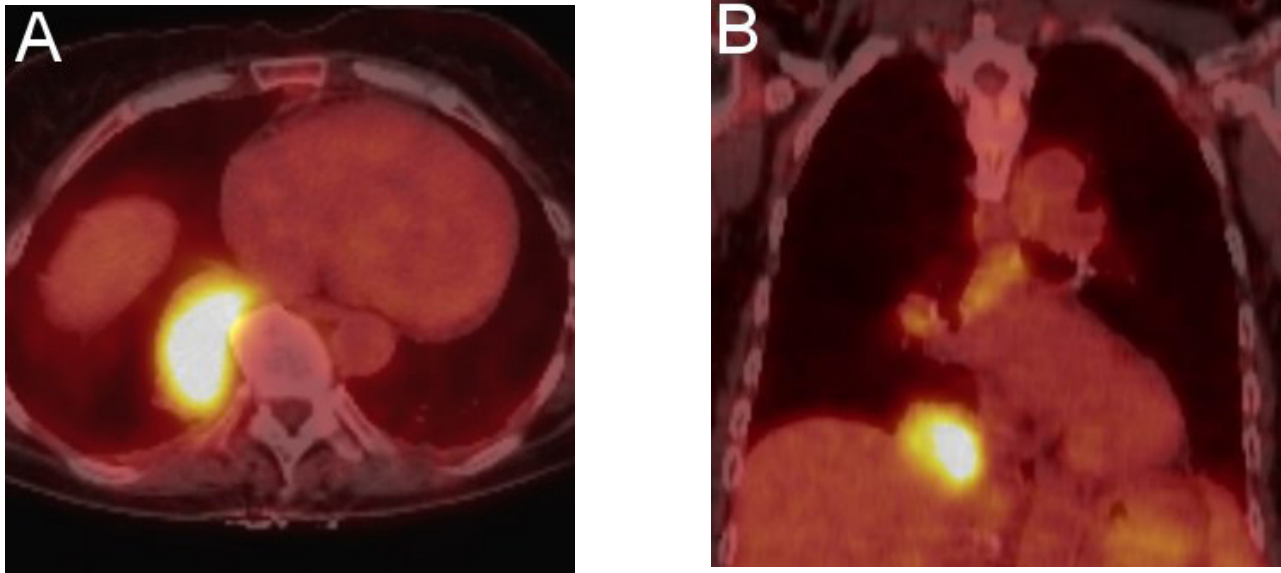
Chest computerized tomography (CT) showed a RLL mass of 5 cm x 4 cm (Figure 1). Also, 18F-fluorodeoxyglucose positron emission tomography (FDG-PET)/CT displayed intense radiotracer uptake in the mass and in non-enlarged subcarinal, right hilar, and mediastinal lymph nodes (Figure 2). Given a high suspicion for malignancy, she underwent mediastinoscopy. Bulky adenopathy



**Figure 1:** Chest CT scan showing RLL mass (4x5cm)

was identified. Biopsies showed no evidence of malignancy or granulomata. Flow cytometry was normal. CT-guided biopsy of the mass was performed and showed necrotic tissue with negative cultures. Due to continued concern for malignancy, video-assisted thoracic surgery was scheduled. Pre-operative laboratory evaluation revealed a creatinine of 2.4 mg/dL (baseline 0.6 mg/dL), which may be due to the contrast used for the CT-guided biopsy. Four days later, creatinine was 5.7 mg/dL. Surgery was deferred, and she was hospitalized. Urine microscopy demonstrated proteinuria, hematuria with dysmorphic red blood cells (RBCs), and RBC casts, consistent with acute glomerulonephritis. Empiric treatment was initiated with pulse steroids. Unfortunately, rapid deterioration in renal function ensued, prompting initiation of plasmapheresis

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**Figure 2.** PET scan showing increased FDG uptake of **A)** RLL mass and **B)** mediastinal lymph nodes

and hemodialysis. Renal biopsy revealed severe crescentic glomerulonephritis with active cellular crescents involving 90% of the viable glomeruli (Figures 3 and 4). Cytoplasmic-ANCA (C-ANCA) was markedly elevated at 11,469 units, which was confirmed by enzyme-linked immunosorbent assay (ELISA) of antibodies to anti-proteinase 3. Rituximab, corticosteroids, and cyclophosphamide were started.

Three weeks after initiation of therapy, C-ANCA titer was 819 units. The patient remained dialysis-dependent for 3 months prior to renal function return. Chest CT at 3 months showed that the mass reduced to 1.5 cm in diameter (Figure 5). Immunotherapy was continued. At one year, repeat C-ANCA titers were negative at 1.4 units and the RLL mass resolved (Figure 6).

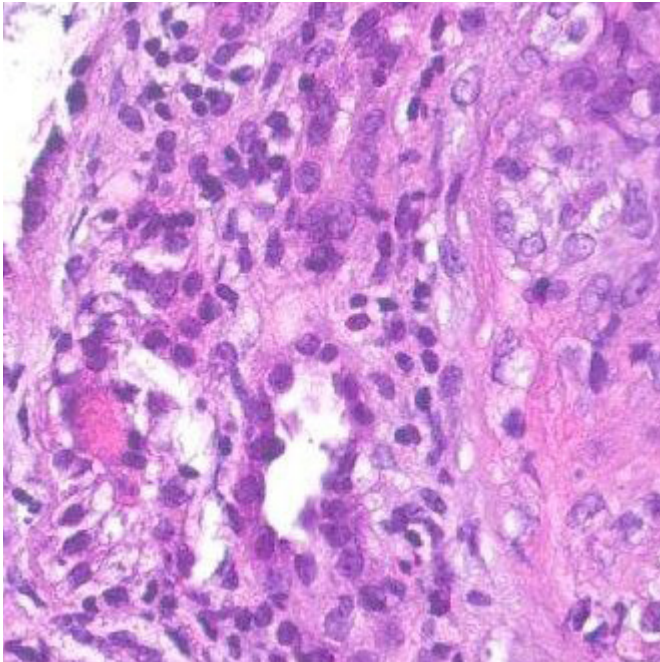
## DISCUSSION

GPA is a small-vessel vasculitis with necrotizing granulomatous inflammation that usually affects the kidneys and the upper and lower respiratory tracts.<sup>7</sup> Approximately 85% of patients develop pulmonary manifestations.<sup>8</sup> Up to half of GPA patients have lower airway abnormalities, including subglottic stenosis, ulcerating tracheobronchitis, and tracheal or bronchial stenosis.<sup>9–10</sup> In 5–45% of cases, diffuse alveolar hemorrhage due to alveolar capillaritis is a prominent feature of pulmonary involvement.<sup>5</sup> Hilar or mediastinal lymphadenopathy is rare. Only ten cases of GPA have been reported with hilar or mediastinal adenopathy.<sup>4, 11–14</sup>

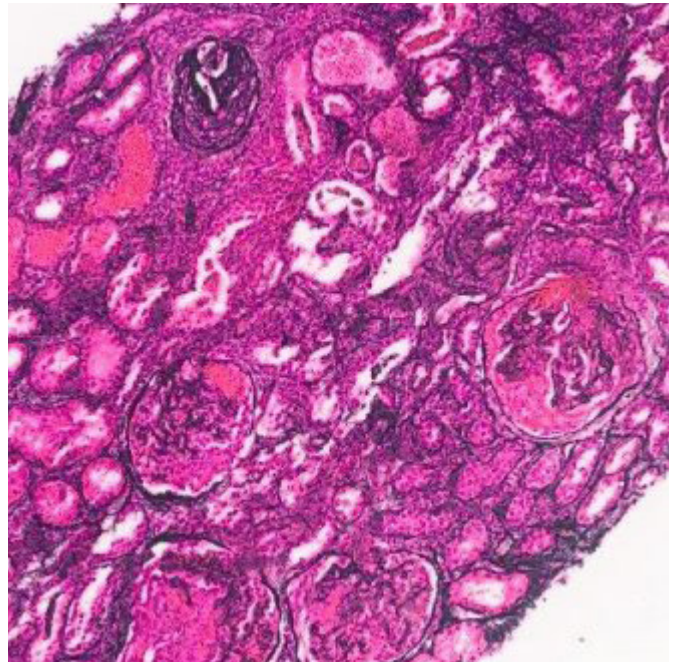
Pulmonary nodules and masses are common radiologic findings, and increased FDG activity has been reported in patients with GPA. The median for the maximum standardized uptake value (SUVmax) is usually within a range of 7.6–9.0, in contrast to our patient who had an SUVmax of 13.1.<sup>15–17</sup> Despite its limitation in differentiating inflammatory diseases from malignancies, FDG-PET/CT could have a role in early diagnosis of GPA. In certain patients, FDG-PET/CT can provide valuable guidance for optimal biopsy site, localization of organ involvement, and characterization of disease severity, which could have treatment implications.<sup>16–19</sup> Furthermore, a case series reported a remarkable decrease in FDG uptake with treatment.<sup>19</sup> This report indicates that FDG uptake correlates with disease activity in GPA.

This case demonstrates an unusual radiological presentation of GPA, with initial findings primarily concerning malignancy, particularly related to the observed increase in FDG uptake in non-enlarged mediastinal and hilar lymph nodes. This unusual presentation resulted in a delay in the diagnosis of GPA, which became evident only with development of acute kidney injury with rapidly progressive glomerulonephritis. Despite this delay, clinical response was achieved with corticosteroids, rituximab, cyclophosphamide, plasmapheresis, and hemodialysis, leading to the recovery of renal function, resolution of CT abnormalities, and normalization of C-ANCA titers.

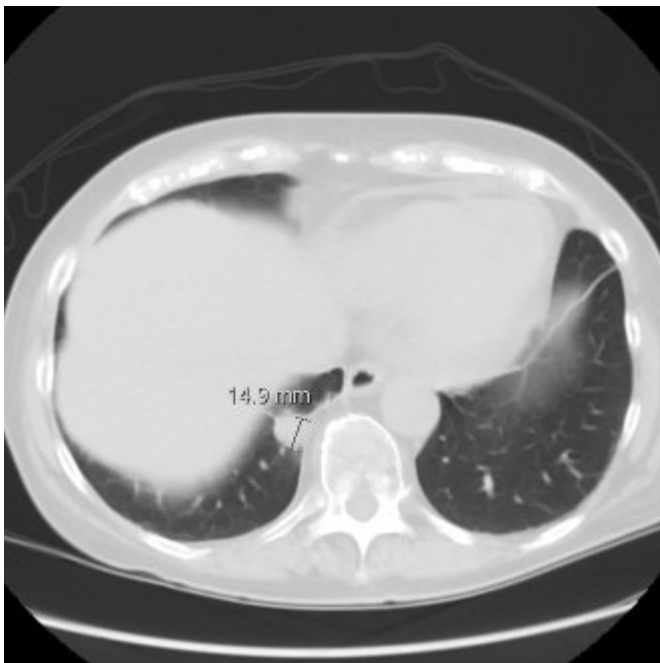




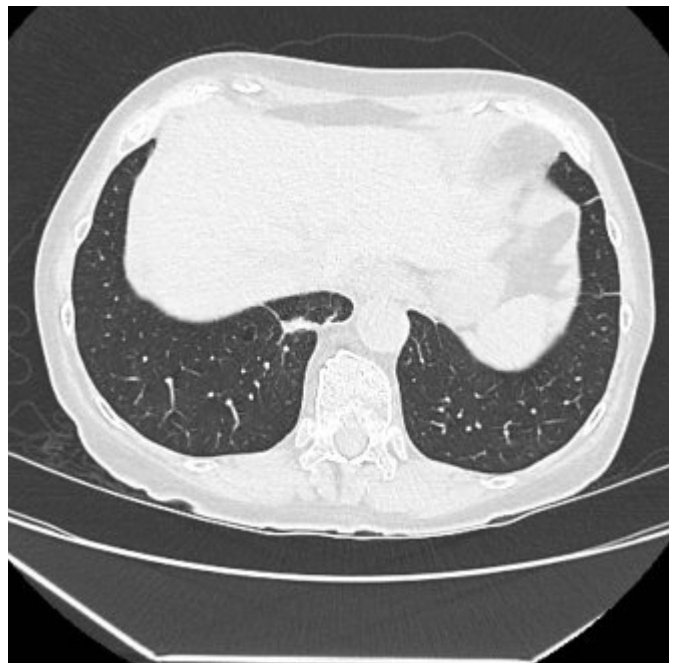
**Figure 3.** Light microscopy of renal biopsy using hematoxylin and eosin stain (H&E stain) showing active cellular crescents within the glomerulus



**Figure 4.** Jones basement membrane stain showing extensive crescentic changes and fibrosis



**Figure 5.** Three month follow up chest CT, showing reduction in mass size



**Figure 6.** One year follow up chest CT, showing resolution of RLL mass with remnant pleural parenchymal scarring

**Conflicts of Interest:** None

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