



A CASE OF IGG4-RELATED INTERSTITIAL LUNG DISEASE MANIFESTING AS RECURRENT NECROTIZING PNEUMONIA

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Introduction:

IgG4-related disease is a newly recognized fibroinflammatory disorder characterized by diffuse lymphoplasmacytic infiltration and IgG4+ plasma cells.

Presentation relies on the primary organ affected. Pulmonary disease can be especially difficult to characterize, as its course may mimic various lung pathologies.

Case Presentation:

33-year old female with PMH of recent Group B Streptococcal bacteremia with aortic valve endocarditis was admitted for severe aortic insufficiency, necessitating aortic valve replacement.

Her course was complicated by acute hypoxic respiratory failure requiring ventilatory support followed by multiple readmissions for right-sided necrotizing pneumonia treated with prolonged courses of antibiotics.

Patient was ultimately re-admitted for left sided chest pain and hypoxia. Initial workup showed elevated inflammatory markers and CT chest with right-sided, multifocal pneumonia and cavitations. Microbiology and immunologic studies were unremarkable.

The patient was again treated empirically with antibiotics but did not improve after 1 month so underwent bronchoalveolar lavage and right lung FNA- results were consistent with IgG4-related ILD.

Treatment was initiated with prednisone 0.6 mg/kg/day and slowly tapered.

Patient was successfully weaned from oxygen support without further addition of DMARDs.

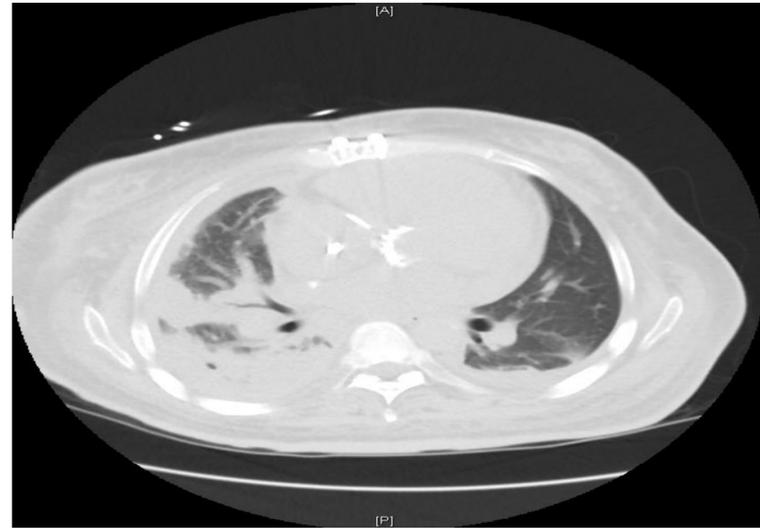


Figure 1. Initial CT Chest demonstrating extensive multifocal pneumonia throughout the right lung with areas of superimposed cavitation/necrosis.

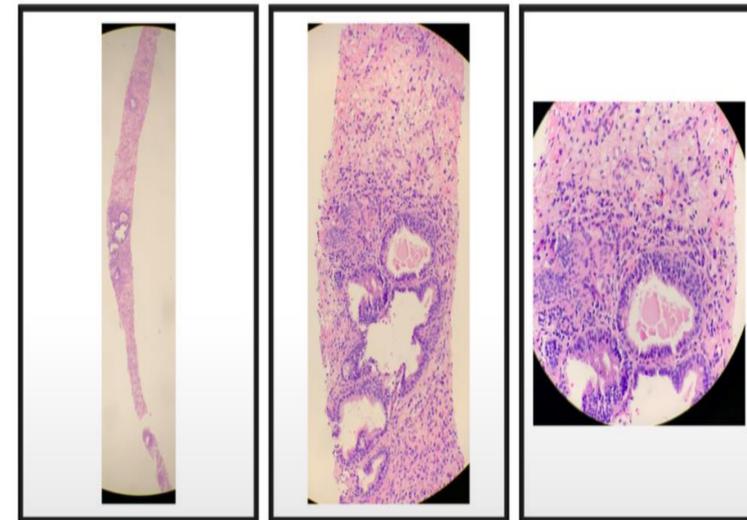


Figure 2. Right Lung FNA H&E stain with interstitial fibrosis, inflammatory lymphoplasmacytic infiltrate.

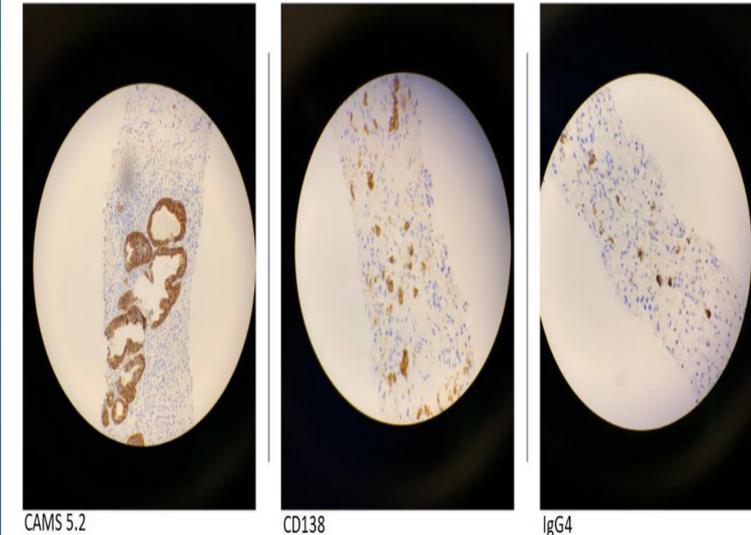


Figure 3. Membrane staining of plasma cells from right lung FNA positive for pankeratin CAM 5.2, CD138, and IgG4.

Discussion:

IgG4-related disease was first recognized in 2003 and is now understood as a spectrum of related diseases.

Pulmonary manifestations may vary depending on site affected and include diverse presentations sharing features of other lung pathologies including pneumonias, cancer, and granulomatous diseases.

Unique to this patient was the recurrent manifestation of necrotizing pneumonia subjecting her to multiple protracted courses of antibiotics that were ineffective.

Treatment required final diagnosis via FNA and the patient was successfully treated with corticosteroids.

Conclusions:

IgG4-related disease encompasses a spectrum of fibroinflammatory disorders that can affect multiple organ systems, including the lungs.

Given the evolving nature of IgG4-related ILD, diagnosis can be especially difficult given the multitude of presentations mimicking other infectious, neoplastic, and inflammatory disease processes.

This case highlights the importances of maintaining a board differential, especially in the setting of ineffective treatments.

Larger studies could further understanding of this complex disease.



Figure 4. Comparison of initial chest X-ray with repeat chest x-ray post therapy demonstrating resolution of pulmonary edema and infiltrates.

References:

1. Stone, John H., et al. "Mechanisms of Disease: IgG4-Related Disease." *The New England Journal of Medicine*, 9 Feb. 2012.
2. Chen, Chiu-Fan et al. "IgG4-related lung disease presenting as interstitial lung disease with bronchiolitis: A case report." *Medicine* vol. 96,49 (2017): e9140. doi:10.1097/MD.0000000000009140
3. Umehara, Hisanori, et al. "Comprehensive diagnostic criteria for IgG4-related disease (IgG4-RD)." *Modern Rheumatology*, vol. 22, no. 1, 2 Jan. 2014, pp. 21-30. Abstract.
4. Campbell, Sabrina., et al. "Clinical review of pulmonary manifestations of IgG4-related disease." *Annals of the American Thoracic Society* vol 11, Issue 9. Sept. 2014.