Medical Image of the Week: Diffuse Alveolar Hemorrhage in a Patient with ANCA Vasculitis and IgG4-Related Disease

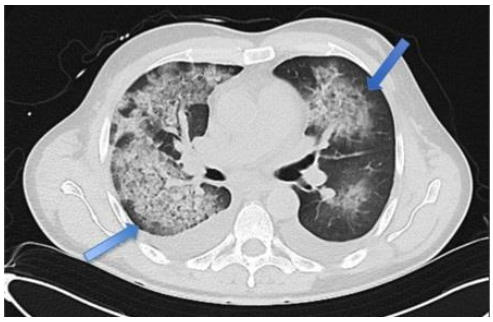

Figure 1. Well-circumscribed, high-density, airspace opacities with a “crazy-paving” pattern in the upper and lower lobes with peripheral sparing (blue arrows) consistent with alveolar hemorrhage.

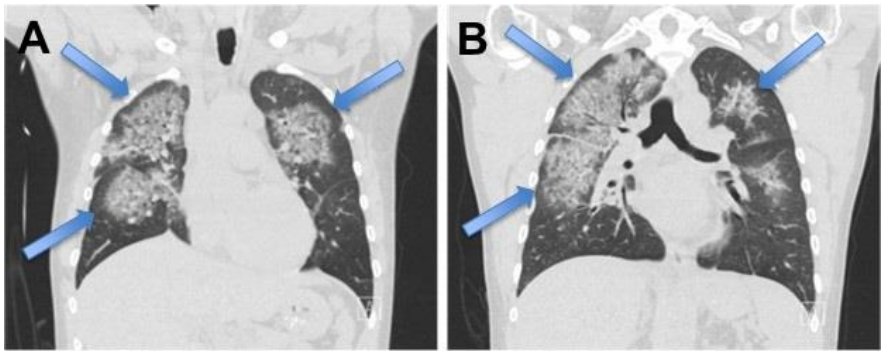

Figure 2. Well-circumscribed, high-density, airspace opacities with a “crazy-paving” pattern in the upper and lower lobes with peripheral sparing (blue arrows) consistent with alveolar hemorrhage.

The patient is a 47-year-old man with a history of bilateral orbital pseudotumor associated with immunoglobulin G4-related disease (IgG4-RD). He presented with progressively worsening exertional dyspnea evolving into multisystemic failure. During the hospitalization, the patient was found to have pauci-immune ANCA-positive vasculitis and glomerulonephritis.

CT images (Figures 1 and 2) show relatively well-circumscribed and extensive upper lung predominant airspace opacities with high attenuation, in some cases with a patchy configuration. A background of interstitial prominence was also noted resulting in a "crazy paving" pattern”, consistent with diffuse alveolar hemorrhage. This was confirmed with bronchoalveolar lavage.

Discussion

IgG4-RD (IgG4 related disease), is an autoimmune condition capable of causing inflammation and fibrosis of multiple organs, most classically the pancreas (1). IgG4 is the least abundant IgG in the serum and the least likely to stimulate immune activation due to its inability to activate complement (2).

The thoracic manifestations that have been described in cases of pure IgG4-RD include solid nodules, which can appear similar to malignant lesions. Interstitial changes have also been described in the form of non-specific interstitial pneumonia pattern, organizing pneumonia, bronchiolitis obliterans, acute interstitial pneumonitis and a sarcoid-like reaction. There may also be pleural involvement and thickening/irregularity of the central airways. The multiple varying presentations and their potential concomitance can lead to misinterpretation of findings (1-2).

This patient presented with the known history of IgG4-RD. The acute symptoms included hemoptysis/diffuse alveolar hemorrhage and renal failure. To the best of our knowledge, pulmonary hemorrhage has not been described as a potential manifestation of this IgG4-RD. Therefore, the later diagnosed concomitant ANCA paucimmune vasculitis, likely explained the observed pulmonary findings. The coexistence of two different autoimmune vasculitides has been described before, both contributing to multiorgan-involvement (3).

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References

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2. Campbell SN, Rubio E, Loschner AL. Clinical review of pulmonary manifestations of IgG4-related disease. Ann Am Thorac Soc. 2014;11(9):1466-75. [CrossRef] [PubMed]
3. Carruthers I, Shingare S, Khosroshahi A, *et al.* IgG4 plasma cell infiltration in granulomatosis with polyangiitis (formerly Wegener's) lung biopsies. 2012 ACR/ARHP Annual Meeting. [Abstract 1534].