Respiratory Distress in Antiphospholipid Antibody Syndrome

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Introduction

Patients with antiphospholipid antibody syndrome experience many pulmonary complications as a course of their disease, most commonly pulmonary embolism or pulmonary hypertension.[1] All serious pulmonary complications should be kept in mind, even without pathognomonic disease features.

Case Description

HPI: A 54-year-old man with triple positive antiphospholipid antibody syndrome anticoagulated with warfarin undergoing a workup for chronic kidney disease and diffuse ground glass opacities on CT presented with acute-onset dizziness, vomiting, and severe dyspnea with exertion after a five hour flight.

Labs:

Chromogenic Anti-Xa 0.19

138	100	39	133
37	28	2.0	

	14		
8.5			182
	41		

Imaging:

- **CT chest**: Multifocal groundglass opacities with septal thickening (Figure 2)
- **Lower extremity dopplers**: Deep vein thromboses in right popliteal and left femoral, popliteal, and gastrocnemius vein
- **Transthoracic echocardiogram:** Normal EF, no left heart strain

Hospital Course:

Patient required 8-10 L oxygen per nasal canula and continued to desaturate with minimal exertion. Bronchoscopy was performed and bronchoalveolar lavage (BAL) was notable for some blood at the end of specimen collection. Patient was started on high dose intravenous steroids, which quickly improved his respiratory symptoms. BAL specimen returned with >95% iron positive macrophages by iron stain (Figure 3), diagnostic for diffuse alveolar hemorrhage (DAH).

Imaging

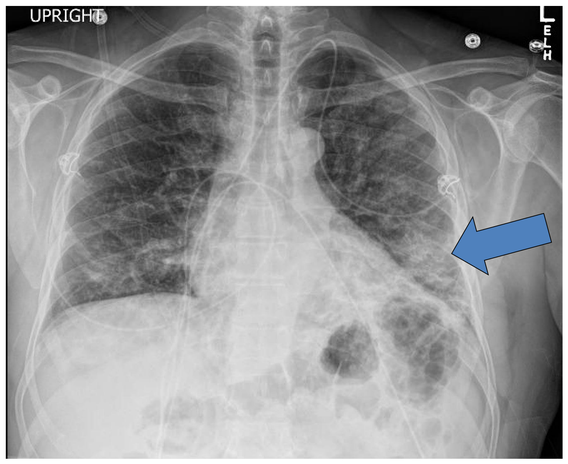


Figure 1: Chest radiograph: Patchy, scattered bilateral groundglass and consolidative opacities, most prominent in the left lower lung (arrow), with mild lateral reticulations.

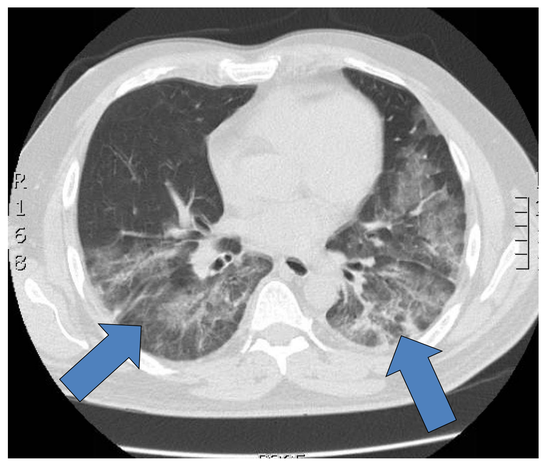
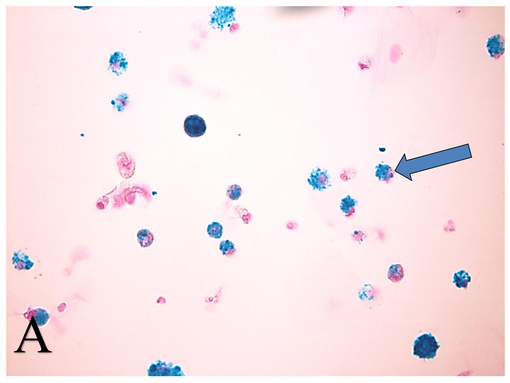


Figure 2: Non-contrast CT showing multifocal groundglass opacities (arrows) with superimposed septal thickening diffusely throughout, left greater than right.

Cytology



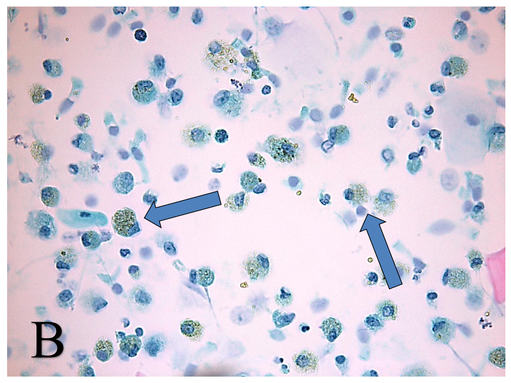


Figure 3:
Bronchoalveolar lavage with approximately 95% of pulmonary macrophages staining positive for iron with Prussian Blue stain (siderophages). **A:** Prussian blue stain, with most of the cells in the field taking up the stain (arrow). **B:** Surepath slide, showing a majority of hemosiderin laden macrophages (arrows).

Clues On Imaging

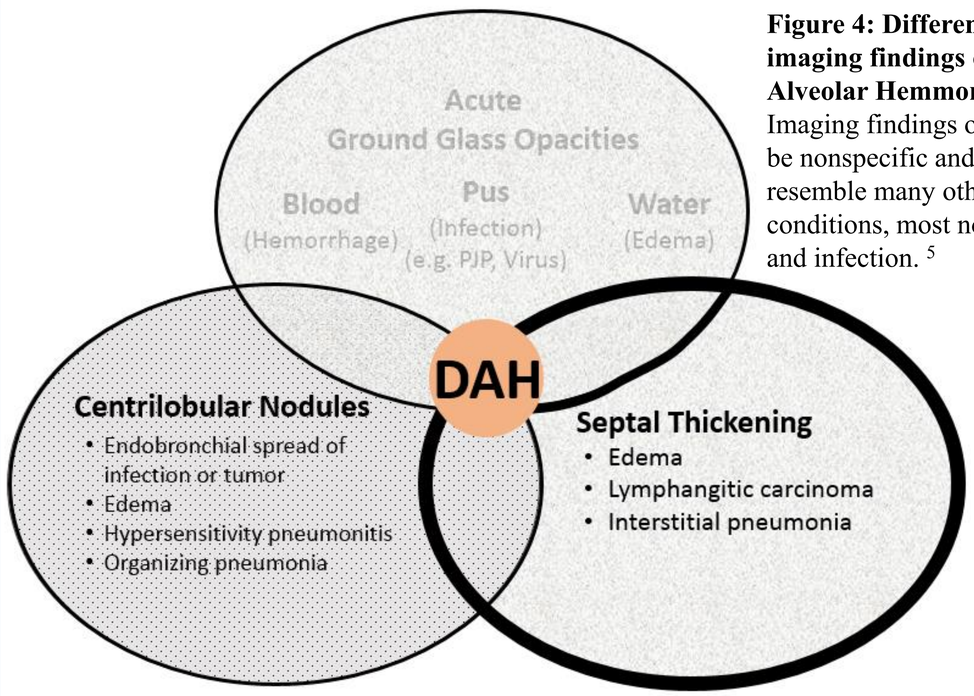


Figure 4: Differential for the imaging findings of Diffuse Alveolar Hemmorrhage
Imaging findings of DAH can be nonspecific and closely resemble many other conditions, most notably edema and infection. [5]

Discussion

Diffuse alveolar hemorrhage (DAH) syndrome occurs when there is damage to the bronchial or pulmonary microcirculation, leading to bleeding into the alveoli.[2] It is defined by a triad of hemoptysis, anemia, and progressive hypoxemic respiratory failure. It is, however, important to note that up to one-third of patients do not experience hemoptysis.[3] Imaging findings concerning for DAH are predominantly central and basilar patchy groundglass opacities and septal thickening,[4] which most broadly resembles edema or infection. BAL with at least 20% siderophages is considered diagnostic (figure 3).[5] DAH can be deadly, particularly if due to capillaritis,[6] as in our patient. The mainstay of treatment is corticosteroids, with or without additional immunosuppressive agents.

Teaching Points

- One–third of patients with DAH do not experience hemoptysis.
- DAH should be suspected in patients with antiphospholipid antibody syndrome with worsening hypoxia, even if hemoptysis is absent.
- Imaging findings may be confused for edema or infection, and steroids should be initiated as soon as infection has been ruled out.

References

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