
Physical examination: Tachycardic, tachypneic, and saturating 91% on room air.

Differential diagnosis: Heart failure exacerbation, bronchitis, pneumonia, bleeding diathesis secondary to anticoagulation, cancer, diffuse alveolar hemorrhage (DAH).

Initial work-up and management on admission:
• Chest CTA: Left upper lobe radiopacity (figure 1).
• Initial laboratory analysis: Hb 9.6g/dl. Cr 2.25g/dl. Elevated ANA titer was 1:320 with homogeneous pattern. A urine drug screening was negative.
• Flexible bronchoscopy: Active bleeding from the left inferior lingula segment (figure 2).
• Descending thoracic aortogram: Normal bronchial vasculature.

Further rheumatology work up:
• Elevated ANCA-MPO > 800 (normal, <1).
• Elevated anti-PR3 27.1 (normal, <1).
• Mildly elevated anti ds-DNA 9 (normal, <5).

Management during admission:
• Treated with intravenous methylprednisolone 100 mg daily for 3 days followed by taper.
• On further assessment of medications, he was treated with hydralazine for at least 3 years prior to presentation (dose range 150-300 mg/day). Therefore, hydralazine was discontinued.
• Discharged home 14 days later in stable conditions.

Case Continued

Figure 1. Chest computed tomography angiogram showing left upper lobe hyperdensity in the coronal plane.

Figure 2. Flexible bronchoscopy showing fresh blood located in the tracheal carina (A) and anterior segment of lingula (B)

Discussion

• We presented an elderly man with DAH in whom ANCA multi-antigenicity (positive MPO and PR3) and extremely elevated ANCA-MPO concentration (800-fold greater than normal) were suggestive of drug induced AAV secondary to hydralazine.
• Overlapping features of SLE, including elevated ANA and ds-DNA titers, and low complement C3 concentration, as seen in this case, are common in hydralazine-AAV.
• Our patient was successfully treated with high-dose corticosteroids and hydralazine discontinuation in the acute setting. Other studies have reported the use of induction therapy agents for idiopathic-AAV, such as cyclophosphamide, rituximab, and plasmapheresis.
• The patient was switched to mycophenolate mofetil for maintenance therapy with no recurrence of disease at 6-month of follow-up, as similarly described in idiopathic-AAV studies.
• Further prospective studies to understand the role of immunsuppressors in drug induced AAV, in addition to offending drug discontinuation, are required.

References