Hemoptysis - A Diagnostic Dilemma

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ATS 2019 Where today's science meets tomorrow's care™
dallas tx MAY 17-MAY 22, 2019

Introduction

- Pulmonary-renal syndrome (PRS) refers to the combination of diffuse alveolar hemorrhage (DAH) and rapidly progressive glomerulonephritis, with DAH being the initial manifestation in up to 50% of the cases.
- Hemoptysis is a common presenting symptom, but may be absent in up to 33% of patients presenting with DAH.
- Furthermore, DAH on presentation is an independent predictor of mortality.
- Microscopic Polyangiitis (MPA) is the most common cause of PRS, but determining the etiology of PRS can pose a diagnostic dilemma. We share our experience through a case of a young female presenting with PRS.

Case Description

A 43-year-old female with a past medical history of Crohn's disease, presented with sudden onset of cough with approximately two cups of hemoptysis. Associated symptoms included nausea without vomiting.
Patient reported experiencing generalized malaise and anorexia for the past one month along with subjective fevers, drenching night sweats, a fifteen pound weight loss and darkening of urine without decrease in quantity. She denied recent travel, exposure to sick contacts or pets at home, cigarette smoke, alcohol consumption or illicit substance use. Crohn's disease was in remission without any medications and she did not have any flare ups in recent years.
On presentation, patient was hypoxic but improved with oxygen supplementation. Vitals were otherwise stable and cardiopulmonary examination was unremarkable.

Lab work revealed microcytic anemia, decreased renal function and absence of coagulopathy. Blood Gas showed mild hypoxia and Urinalysis was significant for moderate hematuria. Chest X-ray revealed bilateral perihilar infiltrates. CT chest without IV contrast showed bilateral multiple nodules with centrilobular distribution. Bronchoscopy with bronchoalveolar lavage revealed progressive bloody return confirming suspicion of DAH. Autoimmune workup revealed ANA+ with speckled pattern, low C3 and C4 levels, positive pANCA, dsDNA and AntiSm Antibodies, with elevated Sm/RNP. Due to high suspicion of RPS, pulse-dose steroid was initiated. Renal biopsy showed Crescenteric glomerulonephritis without scarring, weak immunofluorescence and a component of acute tubular necrosis (Fig 1).
A diagnosis of MPA was made and patient was started on cyclophosphamide. Patient did not have further episodes of hemoptysis during hospital stay.

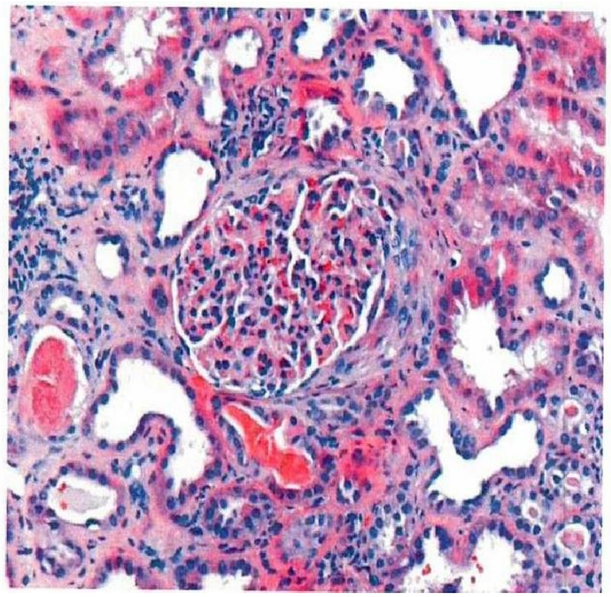

Fig 1. Classic Crescent formation on kidney biopsy under light microscopy

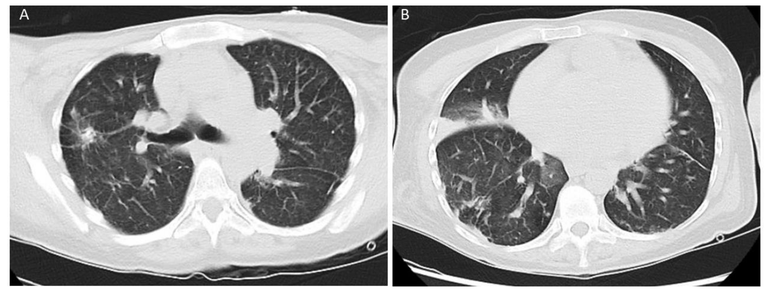

Fig 2. Axial non contrast enhanced CT scan chest, Lung window: showing Dark Brochus Sign (A) and Ground glass opacities (B)

Discussion

- DAH is a potentially life-threatening manifestation of MPA leading to mortality rate of 31%. Factors such as late presentation with hemorrhagic shock, advanced age (>60 years), renal failure, and cardiovascular comorbidity are associated with poor outcomes.
- Therefore, diagnosing MPA requires high index of suspicion and early utilization of invasive diagnostic modalities for prompt management with systemic glucocorticoids, cyclophosphamide and use of plasmapheresis. In advanced cases this management can reduce morbidity and mortality.
- In our patient, pulmonary hemorrhage was not very extensive, and early presentation with prompt initiation of appropriate treatment resulted in good prognosis.

References

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