LEARNING OBJECTIVES

- Recognize that anti-GBM antibody disease is a rare condition.
- Have a high suspicion when appropriate, as early diagnosis and treatment yields better prognosis.
- Understand that some lab findings may lag behind clinical improvement.

CASE PRESENTATION

A 66-year-old man presented to the emergency department with multiple episodes of hemoptysis.

HPI: He reported a sinus infection 3 weeks prior to presentation, followed by fatigue and progressively worsening bilateral knee pain. These symptoms improved until 3 days prior to admission, when he noticed bright red purpuric lesions on his lower extremities, associated with pruritus. He reported dyspnea on exertion and experienced an episode of hematuria within the week of presentation.

PMH: Hyperlipidemia, anxiety, and eczema

Physical Examination: BP: 170/90 mmHg. There was an ulcer on left lateral tongue; a small tender submandibular lymph node, bilateral rhonchi more on the right than left, a palpable purpuric lesions near medial malleolus bilaterally and scattered petechiae (fig f).

Laboratory Data:
- ABG: pH 7.52, PaCO2 28, PaO2 53
- Hgb: 7.8
- Cr: 1.8>5.0
- CRP: 6.2
- ESR: 89
- UA: hematuria

CXR showed “batwing” perihilar pattern (fig c).

CT chest indicated a diffuse alveolar hemorrhage (fig b).

Renal biopsy revealed crescentic lesions in more than 30% of the glomeruli.

HOSPITAL COURSE

- He received high dose steroids, cyclophosphamide, and a series of plasmapheresis treatments with improvement in his condition.
- Anti-GBM antibody trended down to zero from 112 U/ml prior to patient’s discharge.
- Hemoptysis and hematuria resolved within 1-2 weeks.
- Cr level peaked to 5.0 by week 4, plateaued and down trended to his baseline.
- He was in hospital for about 5 weeks prior to discharge with close monitoring of anti-GBM levels by nephrology and primary team.

DISCUSSION

- This patient’s presentation was strongly suggestive of double-positive anti-GBM disease.
- His symptoms were preceded by a sinus infection, hemoptysis, hematuria, and palpable purpura.
- Early diagnosis is vital to the prognosis: c-ANCA positivity improves prognosis.
- The patient’s serum Cr worsened even with clinical improvement: elevated Cr can persist for 3 weeks prior to plateau.
- Compared to using immunosuppressants alone, combined therapy with plasmapheresis, cyclophosphamide, and prednisone prevent further complications and improve mortality.

DOUBLE POSITIVE ANTI-GBM DISEASE

- Anti-glomerular basement membrane (GBM) antibody disease is a rare autoimmune disorder.
- It occurs in less than 2 people per million.
- Circulating antibodies are directed against an antigen intrinsic to the GBM and alveolar basement membrane.
- Symptoms include purpuric lesions, hemoptysis, dyspnea, hematuria.
- Inflammatory markers are elevated.

CXR showed the batwing perihilar pattern.

- Chest CT showed diffuse alveolar hemorrhagic pattern.
- A renal biopsy confirmed the presence of crescentic lesion in greater than 30% of glomeruli.

- Treatment:
  - Consisted of plasmapheresis, cyclophosphamide, and prednisone.
  - The triple therapy is better in preventing complications than immunosuppressants alone.

CONCLUSION

- Anti-GBM antibody disease is a rare and potentially fatal condition with good treatment outcomes when detected early.
- A high index of suspicion is warranted in patients presenting with hemoptysis and hematuria, or with similar laboratory results as seen in our patient.
- Elevated creatinine levels are typical of this disease and may take several weeks to plateau after treatment.

REFERENCES: