Acute renal failure after presentation with FUO
Case study and review

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Initial Presentation

• 80 year old male with history of gout and HTN
• Admitted for cough, fevers, weakness, nausea, mild acute renal dysfunction, and emesis
• One month of cough and nasal drainage treated empirically with levofloxacin and then a Z-pack.
• Progressive anorexia, malaise, cough and then development of fevers to 102 and emesis prompting admission
Prior History

• HTN, gout, impaired glucose tolerance
• Two year history of eustachian tube dysfunction with nasal drainage
  – eustachian tubes placed year prior without improvement in symptoms
Admission Findings

• A&Ox3, appears stated age, tachycardia, dry crackles in bases, no rash, no edema
• T 99.8, P 104, BP 134/72, 96% RA
• T 103.3 later in the day
• WBC 22K: 88% Neutrophils, 4% Bands
• Plt 431K, Hgb 14.2, Na 134, Creat 1.4, Alb 2.9
• CRP 13.3 mg/dl, ESR 100
Initial Workup

- Blood cultures negative
- UA noted trace protein
- Respiratory panel negative
  - Adenovirus, influenza, parainfluenza, RSV, rhinovirus
- CT of chest noted bibasilar peripheral interlobular thickening and 7 3-6mm bilateral pulmonary nodules
- CT of abdomen – no acute process
- CT of sinus noted mild mucosal swelling
Hospital Course

• Initiated on vancomycin and cefepime
• T 103.3 later in the day of admission, then a diurnal fever spike of ~ 101 throughout his hospitalization
• Persistent leukocytosis during hospitalization
• TEE and indium scan negative
• Bone marrow noted hypercellular marrow
• Negative tagged WBC scan
Serologic Tests

• Brucella Ab, West Nile Ab, Q fever Ab, and ricketsia AB negative
• QuantiFERON-Gold TB test – indeterminate
• ANA, SPEP, RF, cold agglutins, CCP – all negative
Day 22 after discharge

- Had been started on prednisone a few days after discharge and felt better with 2 days left of therapy (tapering dose)
- WBC 27K and creatinine 2.0 (up from 1.4)
- Urine analysis noted trace protein and moderate blood (4 RBC’s per HPF)
- Normal blood pressure and relatively normal exam
Second Admission

• 29 days after discharge
• Presented with one day of oliguria and gross hematuria
• Relatively normal physical exam and vitals
• K 5.5, BUN 131, Creat 6.9, Alb 2.7, WBC 17.8K
• Urine analysis – protein and hematuria
• Microscopic exam noted granular casts, dysmorphic RBCs, WBCs
Diagnosis

- Renal biopsy: focal necrotizing glomerulonephritis with crescents and negative for immune or electron dense deposits
- Anti-myeloperoxidase: 331 AU/ml (0-19 negative)
- ANCA associated microscopic polyangiitis (MPA)
Initial Therapy

• Methylprednisolone 500mg IV daily x 3
• Cyclophosphamide 500mg IV x 1
• Prepare for transfer to Benefis for plasmapharesis
• Dialysis x 2 with fluid removal
Evening on Day 2

- Acute shortness of breath
- Hemoptysis
- Non-ST elevation myocardial infarction
Acute SOB with hemoptysis
Benefis Course

• Required only one further dialysis treatment
• Plasmapharesis x 5
• Echo – EF 38%
• Stress nuclear study – mixed infarct and ischemia in LAD distribution
• Discharged home 6 days after transfer
Follow up Course

• Received 4 doses of IV cyclophosphamide
  – Did not tolerate well
  – Given negative anti-MPO AB decision was to proceed to azathioprine

• One year after treatment
  – On azathioprine and off prednisone
  – Creatinine 1.5
  – Follow up echocardiogram noted a normal EF
ANCA-associated vasculitis

- Small vessel disease
- Little to no immune deposits
- MPO-ANCA, PR3-ANCA, ANCA negative
- Microscopic polyangiitis
- Granulomatosis with polyangiitis
- Eosinophillic granulomatosis with polyangiitis
Multi-organ Involvement

• Ear, nose, and throat
• Tracheal and pulmonary disease
• Kidney disease
• Ophthalmic disease
• Cardiac, nervous system, gastrointestinal
• Skin involvement
Case

- Chronic ENT issues over a number of years
- Leukocytosis
- Elevated CRP and ESR (not always the case)
- Rapidly progressive glomerulonephritis
- Pulmonary hemorrhage
- Marked improvement in renal function with aggressive immunosuppression
Question

Of the following medications, which is considered first line maintenance therapy for renal ANCA-associated vasculitis?

A) Mycophenolate mofetil
B) Cyclosporine
C) Etanercept
D) Azathioprine – right answer