1. To recognize the clinical signs of Wegener’s granulomatosis.
2. To understand the importance of early diagnosis and treatment of the disease.

Presentation
• A 17-year-old man presented to the emergency room with a two month history of dry cough, generalized fatigue, fevers and intermittent shortness of breath.

History
• Patient had presented to the ED multiple times in the previous two months with similar complaints and discharged each time with a diagnosis of pneumonia and prescribed antibiotics.
• No other clinically pertinent past medical history.

Physical exam
• Febrile and tachypneic
• Decreased inspiratory phase

Labs
• WBC 11.7
• H&H 8.6/26.8
• c-ANCA positive
• Urinalysis: occult blood and RBC positive

Hospital Course
• Hospital day 3
  • Bronchoscopy revealed alveolar hemorrhage
• Hospital day 4
  • VATS procedure with lung biopsy and chest tube placement
  • Methylprednisolone started for presumptive Wegener’s
• Frozen sections demonstrated organizing pneumonitis
• Hospital day 5
  • Positive c-ANCA
  • Prednisone 1 mg/kg/day started
  • Patient demonstrated rapid improvement in respiratory status and oxygenation

PATHOLOGY

Fig 1. Multiple fibroblastic foci (yellow arrows) indicative of organizing pneumonia.

Fig 2. Capillaritis; a characteristic of Wegener’s. Neutrophils are seen invading the walls of the capillary (green arrows).

Fig 3. Fresh diffuse intra-alveolar hemorrhage (yellow arrows) suggestive of Wegener’s. There are also abundant hemosiderin-laden macrophages (green arrows) to suggest chronicity.

Fig 4. Chest radiographs showing rapid improvement of multifocal alveolar infiltrates after introduction of daily steroids.

Fig 5. CT chest showing multiple dense alveolar lesions following a broncho-vascular distribution (red arrows). This presentation is characteristic for organizing pneumonia.

DISCUSSION
• Wegener’s granulomatosis is a form of systemic vasculitis with an incidence of 10 cases per million people every year and can affect virtually any organ system. Pulmonary findings include nodular opacities on chest x-ray, while lung biopsy demonstrates granulomatous inflammation, geographic necrosis, and vasculitis. A small percentage of patients can present with atypical pulmonary findings.

• This is a case in which Wegener’s granulomatosis presented with pathological findings of an organizing pneumonia and capillaritis.

• In this case, the diagnosis of Wegener’s is made based upon clinical findings, a positive c-ANCA and pathological findings of alveolar hemorrhage and capillaritis.

CLINICAL PEARLS
• Patients with Wegener’s granulomatosis most often present with persistent rhinorrhea, purulent or bloody nasal discharge, oral and nasal ulcers, polyarthralgias, myalgias or sinus pain.

• Some patients may present with symptoms that resemble a common pneumonia that persists for months or weeks and is refractory to antibiotics.

• When Wegener’s granulomatosis is diagnosed, steroid therapy should be initiated immediately as most patients will demonstrate a rapid improvement in symptoms.

REFERENCES