Imaging of Wegener’s Granulomatosis: Pulmonary, Renal, and Sinus Findings

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Outline

• Features of Wegener’s Granulomatosis
• Patient Presentation
• Imaging Findings in Our Patient
  – Pulmonary Involvement
  – Renal Involvement
  – Sinus Involvement
• Our Patient’s Outcome
Wegener’s Granulomatosis

- Multisystemic necrotizing, granulomatous inflammatory process affecting small vessels
- 85-90% associated with c-ANCA (antibody to proteinase-3)

**American College of Rheumatology Diagnostic Criteria:**

2 or more of the following:

1. **Nasal or oral inflammation**: Development of painful or painless oral ulcers or purulent or bloody nasal discharge
2. **Abnormal chest radiograph**: Presence of nodules, fixed infiltrates, or cavities
3. **Urinary sediment**: Microhematuria (>5 red blood cells per high power field) or red cell casts in urine sediment
4. **Granulomatous inflammation on biopsy**: Histologic changes showing granulomatous inflammation within the wall of an artery or in the perivascular or extravascular area (artery or arteriole)

Inflammatory infiltrate surrounding small arteriole, with multinucleated giant cells (arrows).

Robbins and Cotran, 8th ed.
Our Patient’s History

• 18yo male presents with 2 weeks of severe cough, low-grade fever, epistaxis, and maxillary pain
• Past Medical History:
  – Diagnosis of Crohn’s disease 6 weeks ago
  – Psoriasis since infancy
  – Raynaud’s phenomenon
• Family History:
  – Sister and 5 cousins: Crohn’s disease
  – Father: psoriasis and reactive arthritis
  – Paternal grandmother: rheumatoid arthritis
Our Patient’s Physical Exam and Laboratory Abnormalities

• Physical Exam:
  – T 100.3, P 109, BP 127/81, R 16, O2 sat 100%
  – No ulcerations noted in nasal cavity or oropharynx
  – Lungs clear to auscultation in all fields
  – II/VI systolic murmur
  – Psoriatic plaque in right axilla
  – Otherwise unremarkable

• Notable Laboratory Values
  – WBC 18 Urine RBC 53
  – ESR 112 No casts
  – C-ANCA: negative Nitrites negative
  – ANA 1:40 Cr 0.9
  – Blood cultures negative x7, Urine culture negative
  – PPD negative
  – HIV negative
1. Imaging the Lungs in Wegener’s Granulomatosis
Chest X-ray Abnormalities in Wegener’s Granulomatosis

- Discrete focal opacities, nodular or consolidative, typically multiple, rounded or oval, 2-4 cm, with no zonal predilection
- 25% of larger nodules may cavitate, with variable wall thickness, and air-fluid levels if secondarily infected or hemorrhagic

- Patchy or diffuse consolidation, usually secondary to pulmonary hemorrhage
- Wedge-shaped pleural or parenchymal consolidation may represent focal granulomatous change or pneumonia
- Pleural thickening or small effusions may occur
Chest CT Abnormalities in Wegener’s Granulomatosis

- Better seen are discrete focal opacities, nodular or consolidative, typically multiple, rounded or oval, 2-4 cm, with no zonal predilection
- Ground glass halo around nodules may be secondary to hemorrhage
- Pulmonary hemorrhage can appear as diffuse alveolar consolidation
- Tracheal or bronchial wall thickening and subsequent narrowing can be seen

A. Axial CT shows diffuse alveolar hemorrhage with subpleural sparing
B. Sagittal CT shows stenosis of subglottic trachea (arrow)

Ananthakrishnan, AJR 2009
• Our patient had multiple chest X-rays over the course of his hospitalization. Let’s examine the findings.
Our patient’s initial chest X-ray

- PA projection
- Normal cardi mediastinal silhouette
- Subtle infrarhilar opacity
- Lung fields otherwise clear
Progression of X-ray findings (1)

- 10 days after admission
- AP projection
- Note left PICC terminating in upper SVC
- Development of multiple nodular opacities in all lung fields
Progression of X-ray findings (2)

- 13 days after admission
- AP projection
- Interval development of right apical pneumothorax (status post lung biopsy)
- Continued expansion of nodular opacities
- Subpleural consolidations in right middle and lower lung fields, at sites of wedge biopsy
- Mild pulmonary edema

Chest X-ray. PACS, BIDMC.
Progression of X-ray findings (3)

- 18 days after admission
- PA projection
- Right PICC
- Numerous discrete, rounded opacities in both lungs
- Small areas of subpleural wedge-shaped opacity in right middle and lower lung fields, at sites of wedge biopsy
- Possible new cavitary lesion

Chest X-ray. PACS, BIDMC.
Our patient also had two chest CTs during his hospital course. Let’s examine the findings to better characterize his disease.
Our patient’s initial chest CT (1)

- Central right middle lobe opacity with irregular borders and ground glass at periphery
- Multiple small nodules
Our patient’s initial chest CT (2)

- Cavitation in nodule in left lower lobe
- Small nodules in other lung fields, some solid and some ground glass
Our patient’s initial chest CT (3)

- Multiple round nodules of varying sizes in central and subpleural zones
Our patient’s follow-up Chest CT (1)

- Enlargement of right middle lobe opacity
- Ground glass haze posteriorly
- Enlargement of smaller nodule
Our patient’s follow-up Chest CT (2)

- Evolution of multiple round nodules, some well-defined and some hazy, of varying sizes in multiple lobes and zones of the lung
- Cavitation no longer seen in left lower lobe nodule
Our patient’s follow-up Chest CT (3)

- Multiple intraparenchymal and subpleural nodules, some solid and some with ground glass character
- Slight pleural thickening and atelectasis, particularly in right lower lobe posteriorly
• Given these findings, what entities are high on our differential diagnosis?
Differential diagnosis for lung findings

• Rapid development of multifocal discrete nodular opacities in all lung fields, some with central cavitation
  – Infection: septic emboli, abscesses, tuberculosis, fungal infection
  – Inflammation: autoimmune diseases including vasculitis, sarcoidosis, rheumatoid nodules
  – Metastases: can be similar in appearance, but less likely given demographics of patient and rapidity of progression
• Our patient had a wedge lung biopsy midway through his hospital course, which showed granulomatous inflammation with multiple areas of vasculitic involvement, consistent with the diagnosis of Wegener’s granulomatosis.
• Multiple cultures and stains were negative for infection.
2. Imaging the Kidneys in Wegener’s Granulomatosis
Renal findings in Wegener’s Granulomatosis

• Ultrasound:
  – Typically shows non-specifically increased echogenicity without specific Doppler abnormalities
  – Rarely, may show one or more infiltrative mass-like lesions (Carazo 2001)

• CT:
  – Affected kidney may show reduced nephrogram after contrast administration
  – Rare mass-like lesions reported to be ill-defined and hypointense; may have thin perfusing cortical rim

• MRI:
  – Infiltrative lesions may be isointense on T1 and show heterogeneous T2 intensity
• Our patient had an abdominal CT early in his hospital stay to assess for abscess as a cause of his fever. No abscess was found, but let’s examine his kidneys.
Abdominal CT

- Lower pole of right kidney: ill-defined hypodensity in nephrographic phase of contrast administration
Abdominal CT

Lower pole of right kidney: ill-defined hypodensity in nephrographic phase of contrast administration

Coronal Abdominal CT on soft tissue windows. PACS, BIDMC.
Renal ultrasound

• Doppler images: hypovascularity of right lower pole

Sagittal ultrasound of right kidney. PACS, BIDMC.
• Given these findings, what is our differential diagnosis for this renal abnormality?
Differential diagnosis for renal lesion

• Ill-defined, hypodense, hypovascular lower pole lesion:
  – Focal pyelonephritis, abscess
  – Vascular disruption: infarct, vasculitis
  – Neoplasm could have identical appearance on CT, but less likely in this case since malignancies are typically hypervascular
• This renal lesion was not biopsied, so its etiology is still not certain, though it is hypothesized to be related to vasculitic involvement of the kidney.
• Fortunately, he did not progress from microhematuria to fulminant glomerulonephritis. His creatinine remained stable.
3. Imaging the Sinuses in Wegener’s Granulomatosis
Sinus findings in Wegener’s granulomatosis

• Early CT changes:
  – Non-specific mucosal thickening and antral opacification
• Later CT changes:
  – Opacification of paranasal sinuses
  – Nasal septal thinning
  – Bony destruction
• Early MRI changes: hyperintense T2 signal in inflammatory soft tissue
• Later MRI findings: hypointense T2 signal in inflammatory soft tissue
Other Head and Neck findings in Wegener’s Granulomatosis

• Orbital involvement:
  – CT: Ill-defined soft-tissue mass obscuring optic nerve and extraocular muscles with bony destruction
  – MRI: low to intermediate T1 and T2 signal

• Middle ear and skull base:
  – CT: Opacification of middle ear and mastoid air cells

• CNS involvement:
  – MRI: Non-specific high T2 or FLAIR signal may be seen on MRI related to small vessel vasculitic change
• Our patient had a CT of the sinuses to assess for disease given his epistaxis and facial pain. Let’s examine the findings.
CT of the sinuses in our patient

- Fullness of anterior nasal soft tissues, particularly on right, without definite polyp
- Minimal mucosal thickening of left maxillary sinus

Axial CT of the sinuses on bone windows (A) and soft tissue windows (B). PACS, BIDMC
CT of the sinuses

- Thickened nasal mucosa, particularly on right
- Minimal mucosal thickening of left maxillary sinus

Coronal CT of the sinuses on bone windows (A) and soft tissue windows (B). PACS, BIDMC
• Though not definitively related to Wegener’s granulomatosis, these non-specific findings of mucosal thickening in the nasopharynx and maxillary sinus could correspond to early changes of the disease.
Our patient’s outcome

• Our patient’s microhematuria and epistaxis, in conjunction with suggestive chest imaging and biopsy, were consistent with a new diagnosis of ANCA-negative Wegener’s granulomatosis by American College of Rheumatology criteria.

• Our patient was discharged on day 18 of hospitalization after improvement of symptoms on steroid therapy. He will follow up with cyclophosphamide treatment as an outpatient.
Thank you

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