Cystic Pneumocystis Pneumonia

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Patient Presentation

• 43 y.o. male with AIDS presents with 1-month h/o fatigue, non-productive cough, dyspnea; hemoptysis on the day of admission.

  – Last CD4=9 cells/mm³. Viral load=67,000
  – Off HIV meds x 1 month
  – Off PCP prophylaxis (pentamidine) x 2 months
PMH

- HIV$^+$ since 1993
  - PCP x 2
  - Oral thrush
  - Toxoplasmosis (1995)
  - Pneumococcal Sepsis
- HepB $^+$, HepC $^+$

Social history:
- 40 pack-year smoking history
- No IVDA
Requisition from Primary Care Physician

- Physical exam findings:
  - Decreased breath sounds at L base
  - Rales on L side

- Please obtain CXR to R/O pulmonary process
Patient Initial Presentation

6/20/02

Consolidation
Air-bronchogram

Multiple cystic lesions

Courtesy of James Busch, M.D. and Matt Spencer, M.D.
Patient Initial Presentation

Courtesy of James Busch, M.D. and Matt Spencer, M.D.
Comparison with Prior Study

8/15/00

6/20/02
Helical CT on Presentation

Cysts
Variable size, wall thickness (2-7 mm)

6/21/02 No Contrast
Helical CT on Presentation

- Patchy ground-glass opacity
- Cystic lesion
- Cavitary nodule
Differential Diagnosis for Our Patient Includes:

- Atypical Pneumocystis pneumonia
- Mycobacterial pneumonia
- Fungal pneumonia
  - Cryptococcus, aspergillosis
- Malignancy
  - KHSV, lymphoma
- Bacterial pneumonia
- Multiorganism infection
Diagnosis

• Patient was admitted for further workup and treatment of suspected pulmonary infection.
  – All cultures were negative
  – Three induced-sputums all negative for PCP, AFB

• Nonetheless, given clinical and radiological suspicion, cystic PCP is the most-likely diagnosis

• Empiric treatment was initiated
  • IV pentamidine 4 mg/kg/day x 21 days
  • Follow clinical and radiological response to therapy
Subsequent Study Demonstrates Improvement

6/20/02

7/27/02
PCP and AIDS

- Most common life-threatening pulmonary infection in HIV\(^+\) patients
  - CD4 T-Cell count < 200 cells/mm\(^3\)
  - Declining incidence with improved prophylaxis (TMP-SMX).
- Increased frequency of non-classical manifestations.
PCP Signs and Symptoms

- Fever
- Malaise
- Shortness of Breath
- Non-productive Cough (30% productive)

Onset:
- Signs/symptoms develop slowly in HIV+ patients and are often present for weeks-months prior to presentation.
- Course is more acute for most non-HIV+ immuno-suppressed patients, with signs/symptoms present on average for 3-5 days prior to presentation.

Typical Findings

• Elevated LDH
  – high sensitivity, low specificity
• Arterial hypoxemia
• Widened alveolar-arterial $O_2$ gradient
• Decreased CO diffusing capacity
• Diffuse bilateral PNA on plain radiograph

Diagnosis

• Requires detection of organism
  – Induced sputum
  – Bronchoalveolar lavage
  – Biopsy

• Does not grow in culture
  – Stains, immunohistochemistry, PCR

• Patients with typical clinical and radiologic findings are often treated empirically
  • Radiographic abnormalities often clear within weeks of therapy

Pneumocystis

Cysts filled with sporozoites

Trophozoite

Giemsa

Methenamine

From: http://pathhsw5m54.ucsf.edu/overview/fungi1.html
Classic CXR Features

Up to 39% may have normal CXR! (Boiselle, 1999)

Bilateral, diffuse symmetric consolidation

Courtesy of Phillip Boiselle, M.D.
Classic CT Features

Ground-glass opacity
(intra-alveolar exudate)

Patchy, geographic distribution with central predilection

Courtesy of Phillip Boiselle, M.D.
Foamy Eosinophilic Exudate

Typical pattern: fibrin, dead cells, cysts + trophozoites

From: http://pathhsw5m54.ucsf.edu/overview/fungi1.html
Changing PCP patterns in HIV

• Increased prevalence of cystic disease

• Increased prevalence of spontaneous ptx

• Upper lobe distribution of parenchymal opacities
  • Previously thought to be found mostly in setting of aerosolized pentamidine therapy.

Broadening Spectrum of Abnormalities

- Lung nodules and masses
  - Often represent granulomas
  - May undergo cavitation
- Lobar consolidation
  - 11% PCP cases vs. 61% of bacterial PNA (Boiselle, 1999)
- Bronchiolitis obliterans.
- Interstitial fibrosis
Cystic Pneumocystis Disease

- Cysts vary in size, shape, number, wall thickness
  - Thin-walled (<3mm), usually air-filled
  - Pathophysiology of formation uncertain
  - Usually multiple, bilateral
  - May be intraparenchymal or subpleural
  - Upper lobe predominance

- Cystic disease now occurs in 10-34% PCP cases
- Cysts in HIV patient are highly suggestive of PCP

Cystic PCP and Pneumothorax

- Ptx was diagnosed in 35% HIV+ patients with cystic PCP vs 7% with non-cystic PCP
- Spontaneous PTX is virtually diagnostic of PCP in an HIV+ patient.
- Ptx secondary to PCP infection is associated significantly with increased morbidity and mortality
  - Ptx in cystic PCP are often refractory to chest tube drainage and may require pleurodesis or surgery.

Cystic PCP & Pneumothorax

Numerous cysts, variable size + wall thickness

PTX

Coronal maximum intensity projection (MIP)

Courtesy of Phillip Boiselle, M.D.
Cystic PCP and Pneumothorax

- Ipsilateral interspace widening
- Contralateral mediastinal shift
- PTX
- Left basilar cysts

Courtesy of Phillip Boiselle, M.D.
Upper Lobe Distribution of Cysts

PA radiograph of patient with upper lobe pattern

CT slice through upper lobe

Courtesy of Phillip Boiselle, M.D.
Disseminated Pneumocystis Infection Remains Rare

- Multiple areas of low attenuation
- Micro-calcifications (granulomatous response)

Courtesy of Herbert Gramm, M.D.
Conclusions

- PCP remains the most common life-threatening infection of HIV\(^+\) patients
- Diagnosis requires detection of organism
  - HIV\(^+\) patients are often treated empirically if there is sufficient clinical and radiological suspicion
- Radiologic patterns of PCP are changing
  - Increased prevalence of disease with cysts, ptx, and upper lobe predominance
- Suspect PCP in HIV\(^+\) patient that presents with cystic disease or ptx
References

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