

HOLY C.O.W.!

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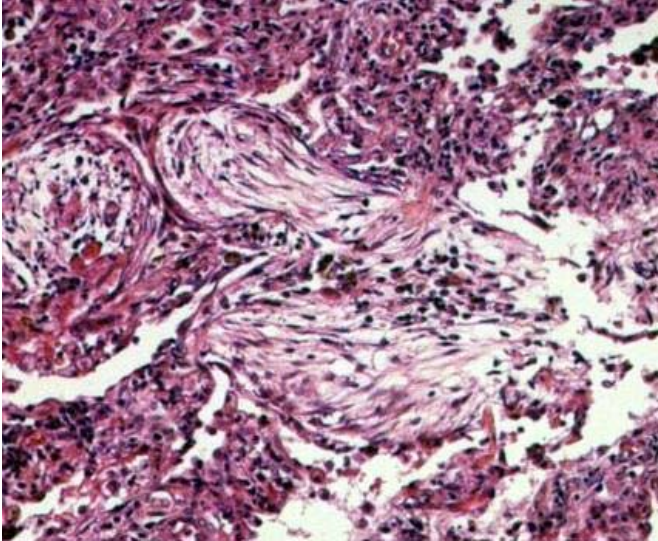
Clinical Question of the Week #7
August 11th, 2008 through August 18th,
2008

Please e-mail your answers to Kuo, Tim, Wendy, and Kevin (klian@mednet.ucla.edu; tprovias@mednet.ucla.edu; wsimon@mednet.ucla.edu; kbreger@mednet.ucla.edu) by 0800 on Monday, August 18th, 2008. The resident or intern with the most correct answers at the end of each month will receive a prize!

Case: A 57-year-old man presents with three weeks of worsening cough and dyspnea. His initial symptoms included a flu-like syndrome with fevers and malaise, followed by decreased appetite and an eight pound weight loss over the same period of time. His past medical history is notable for hypertension and BPH; he is a non-smoker and works at a law firm. Examination reveals bilateral inspiratory crackles, and labs reveal an elevated WBC with neutrophil predominance. A CXR and HRCT chest were obtained and are shown below. Additionally, the patient later had a lung biopsy, also shown below.



CXR and CT chest



Lung Biopsy

Questions:

1. What is the diagnosis?

Cryptogenic organizing pneumonia (COP, also known as idiopathic bronchiolitis obliterans organizing pneumonia or idiopathic BOOP) is a proliferative bronchiolitis of unknown etiology. Other cases of organizing pneumonia (proliferative bronchiolitis) may be seen in association with connective tissue diseases, malignancy, drugs and other interstitial pneumonias. Incidence has been estimated at ~6/100,000, with onset usually in the fifth to sixth decade of life. Men and women are affected equally and smoking is not a contributing factor.

Patients usually present subacutely (with symptoms for less than two months in most patients at the time of presentation), though about half of patients will experience an onset similar to the flu with fever, malaise, fatigue, and cough. Most common features include persistent nonproductive cough, dyspnea with exertion, and weight loss. Exam reveals bilateral rales; wheezing is rare and clubbing is usually absent. Laboratory evaluation is often nonspecific, with elevated WBC, ESR, and CRP. PFTs will reveal mild to moderate restrictive defect and decreased DLCO; resting or exercise-induced hypoxemia is common. BAL reveals foamy macrophages, decreased CD4/CD8 cell ratio, but increased T cell activation.

Lung biopsy is usually needed to confirm diagnosis, although migratory bilateral infiltrates on CT chest can be suggestive (as below). With treatment with steroids (see below), recovery occurs in approximately 2/3 of patients over weeks to months. Recurrence is common and may require long term immunosuppression. Overall prognosis is usually good. (1)

2. What is notable about the pattern of infiltrates seen on radiology imaging in this disease?

Features seen on chest imaging include patchy areas of consolidation, ground glass opacities, often with a more peripheral location, but frank consolidation with air bronchograms are also seen. Wedge shaped infiltrates can be seen on CT chest, with the base along the pleural surfaces, and are usually found in the lower lobes. Honeycombing is rare and usually seen as a late manifestation of progressive disease. A classic feature is migration of consolidated areas on CT chest over time. A less often seen feature is linear opacities on CXR and CT, with an interstitial pneumonitis pattern,

which is associated with a poorer prognosis. (0.5 for migratory consolidation, 0.5 for other radiologic finding)

3. What pathologic feature is shown on the biopsy specimen?

Masson bodies, characteristic intraluminal buds of granulation tissue and fibroblasts, are seen in the biopsy above and are located in the respiratory bronchioles, alveolar ducts, and alveoli. Inflammatory changes are also seen in the alveolar walls, with foamy macrophages in the alveolar spaces. (0.5)

4. What is the treatment?

Corticosteroids are the mainstay of therapy for COP, with usual starting dose of 1mg/kg and continued for 4-8 weeks with subsequent taper. Other immunosuppressive agents have been used, including cyclophosphamide. Some reports of response with macrolide antibiotics have been made. Finally, in focal organizing pneumonia, resection is performed. (0.5)