Approach to Nonneoplastic lung disease, especially ILD

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Course Description

5 part primer on the C-R-P approach to the diagnosis of "interstitial lung disease", (or "ILD").

Intended audience:

Pulmonologists, Radiologists, and Pathologists (and their trainees)



Course Description

Part 1: Introduction to Domains and Patterns in ILD: Focus on <u>acute</u> disease

Part 2: Additional patterns of lung fibrosis and inflammatory infiltrates in ILD.

Part 3: Diffuse lung diseases with granulomatous features.

Part 4: ILD with airway-centering and bronchiolitis.

Part 5: Non-neoplastic lung disease potpourri



Non-neoplastic Lung Pathology III

Diffuse lung diseases with granulomatous features

KO LESLIE MD COMMENTARY: TV COLBY MD





Case Presentation

What constitutes a "granuloma" in lung?

Granulomatous lung disease prototype—Sarcoidosis

Common and uncommon causes of granulomatous lung disease

Optimal approach to granulomas in a lung biopsy



How granulomatous diseases present on imaging

Morphologic spectrum:

Localized solitary lesion v. diffuse ILD

Large v. small

Well formed v. poorly formed

How host immunity influences granulomas



Clinical Case

A 50 year old Caucasian woman was in her usual state of good health when she developed a mild upper respiratory tract infection.

During the recovery phase of that illness, she was bothered by persistent bronchitis with a mildly productive cough.



On advise from her physician husband, she began self treatment with hot moisturized air, while using an indoor hot water soaking tub ("Hot tub" of "Jacuzzi").

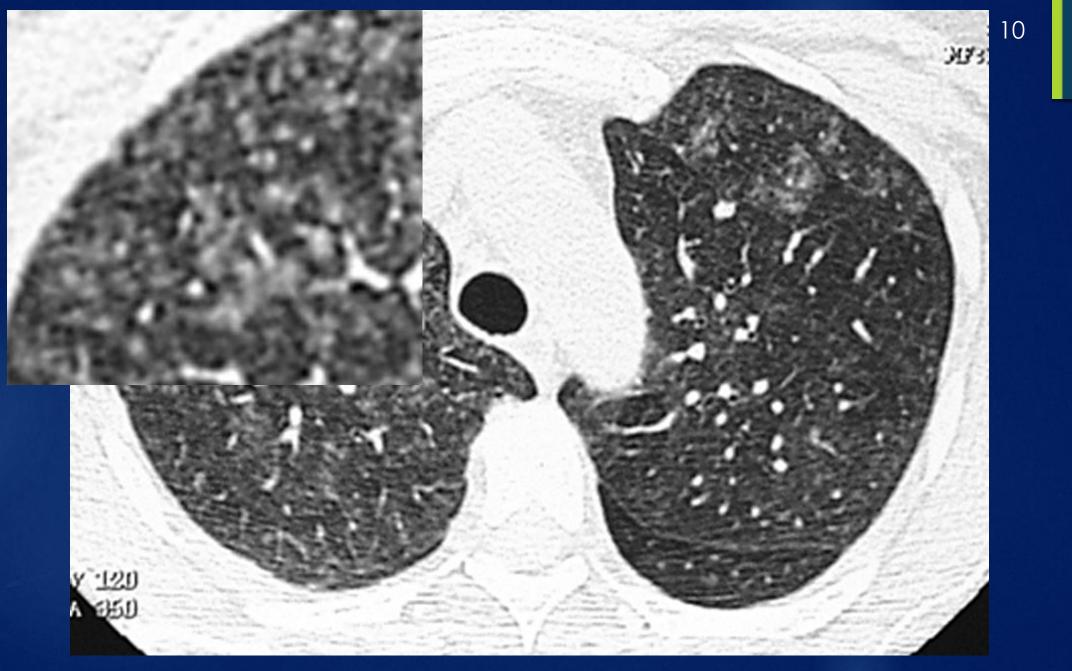
Daily, she would place a towel over her head, and breathe in deeply while seated in the tub. Over the next few weeks, her cough persisted and was progressively accompanied by shortness of breath.



Clinical Case

Eventually she was seen by a pulmonary specialist and a CT scan was performed...

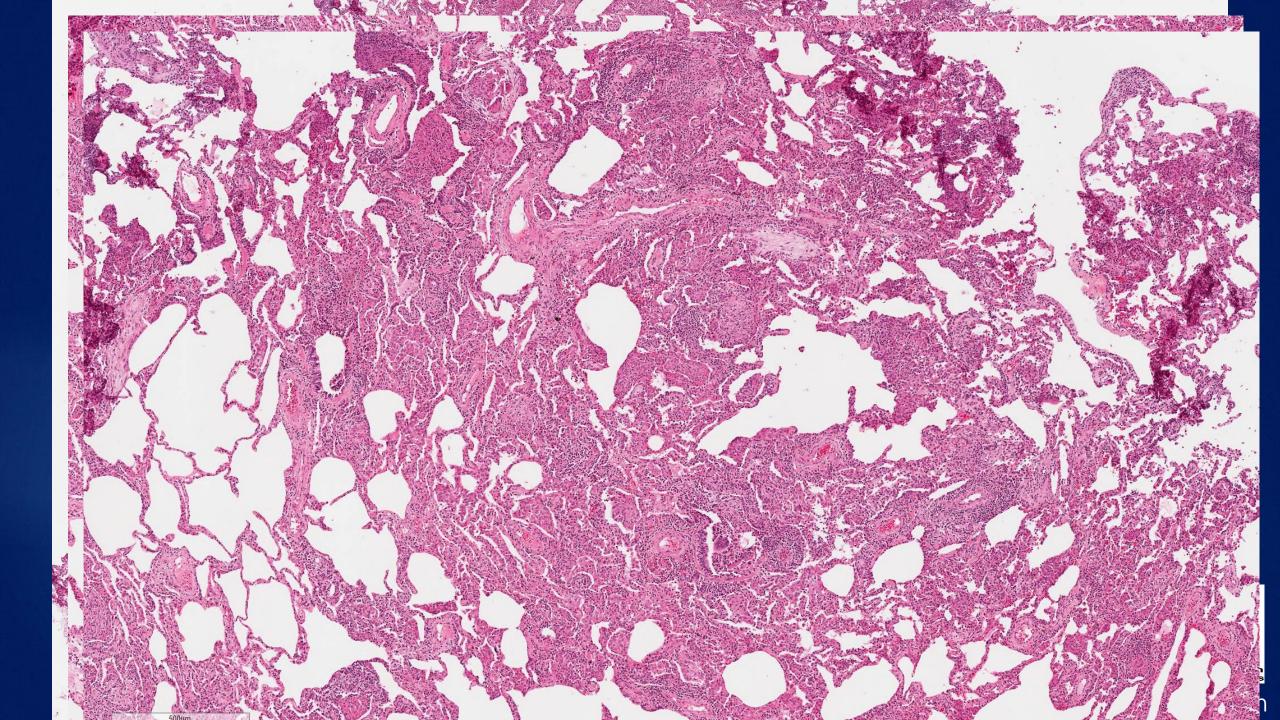


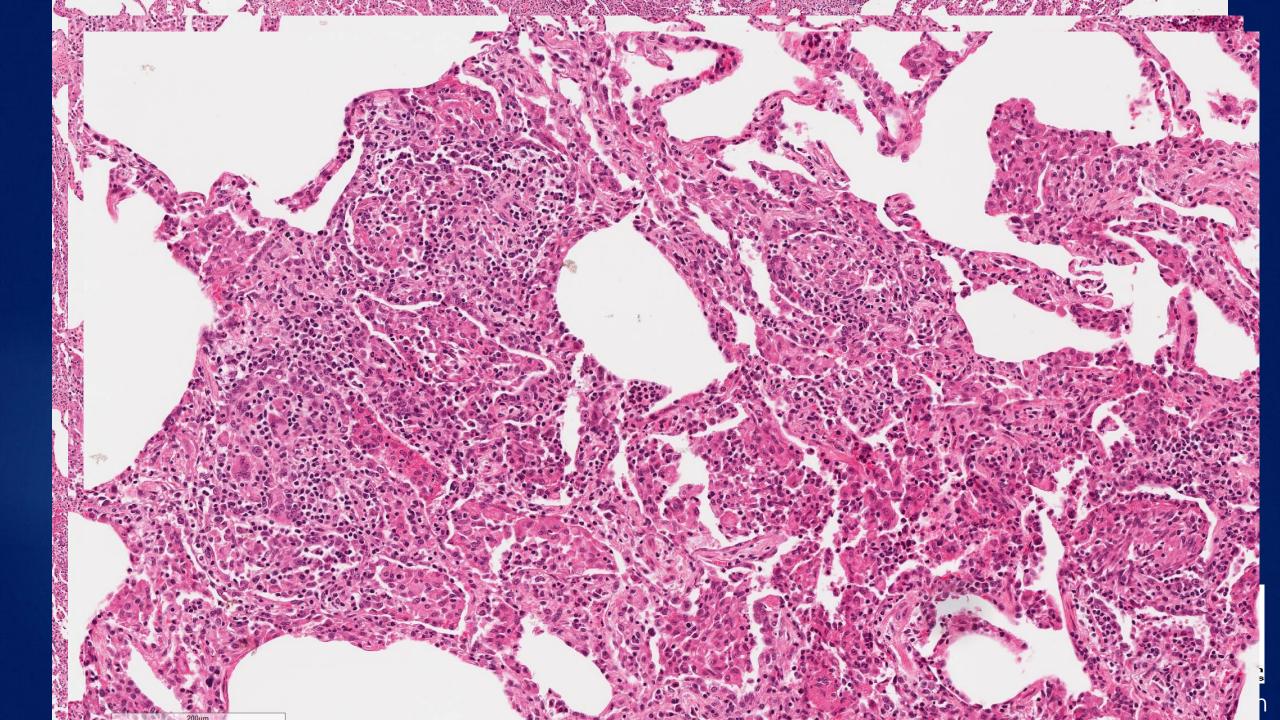


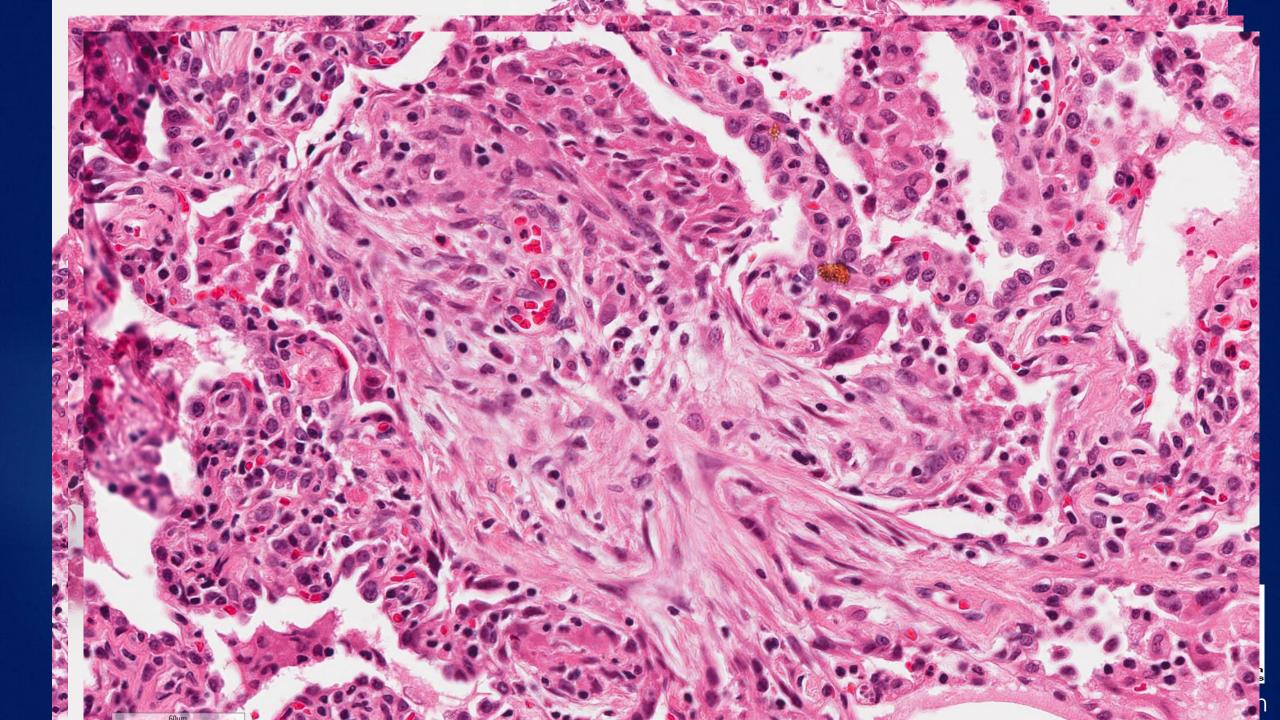
LungPath

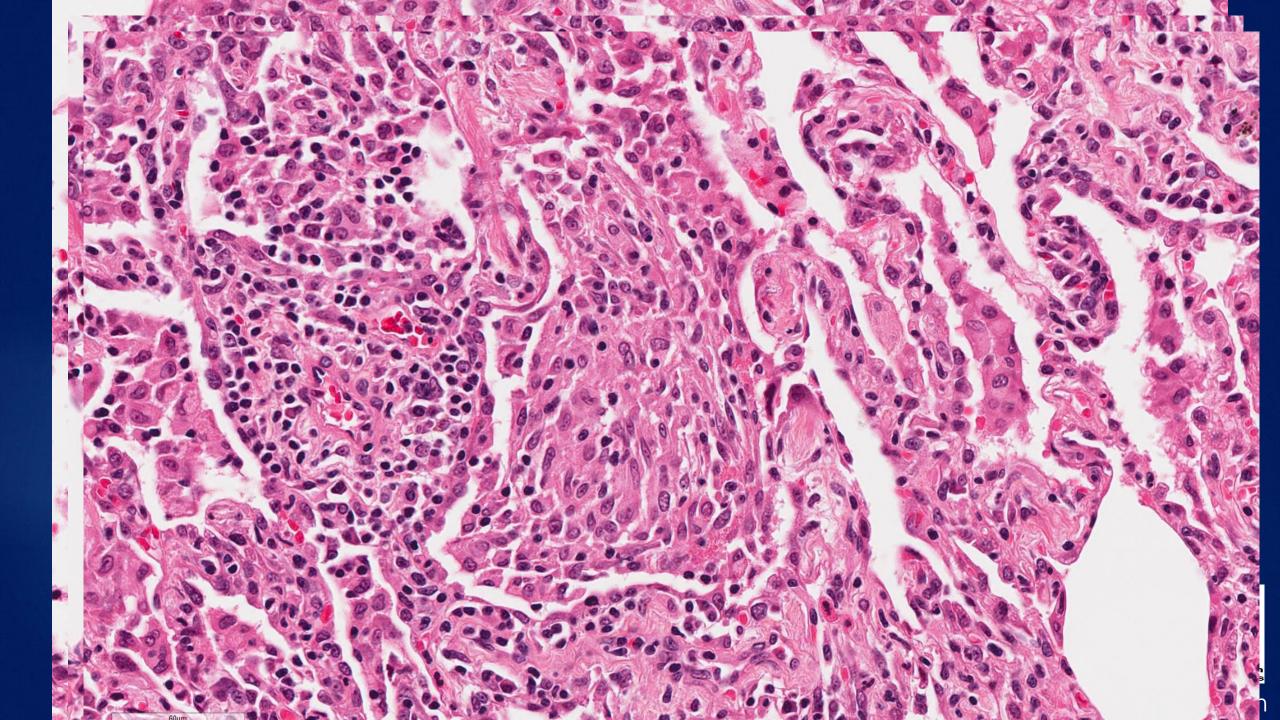
...and the SLB shows

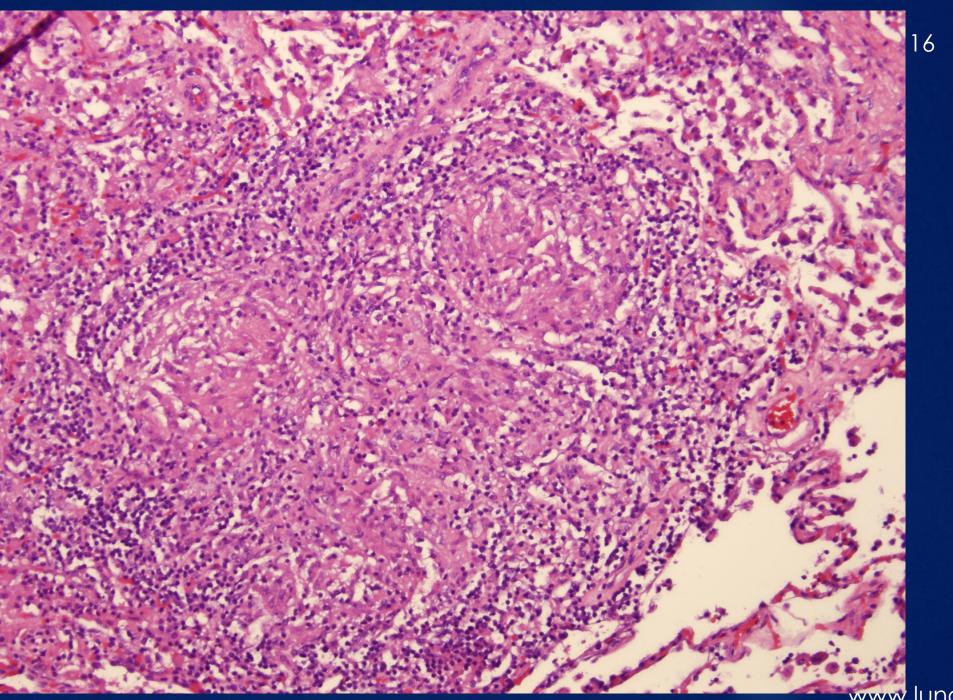




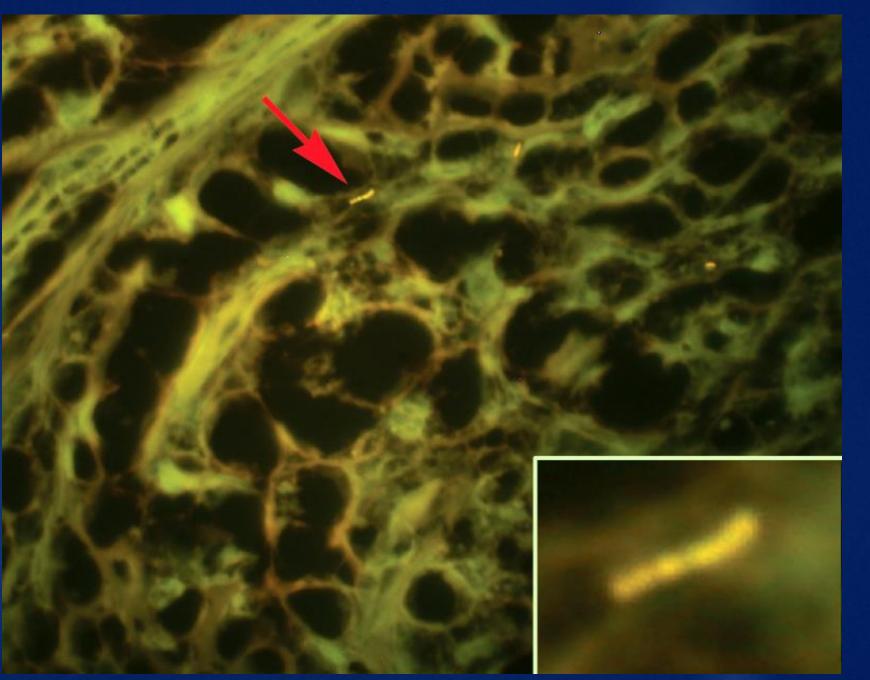
















Diffuse granulomatous pneumonitis resulting from bioaerosol inhalation of M. Avium (so-called "Hot Tub Lung")

Case Analysis

Four Domains

Clinical/lab presentation Radiologic findings Pathologic injury pattern Disease entity that fits Slowly progressive dyspnea Cough, Hot Tub use Bilateral ill-defined centrilobular nodules, upper lung zones

Interstitial chronic inflammation, small nonnecrotizing granulomas, acid fast organisms

Hot Tub Lung

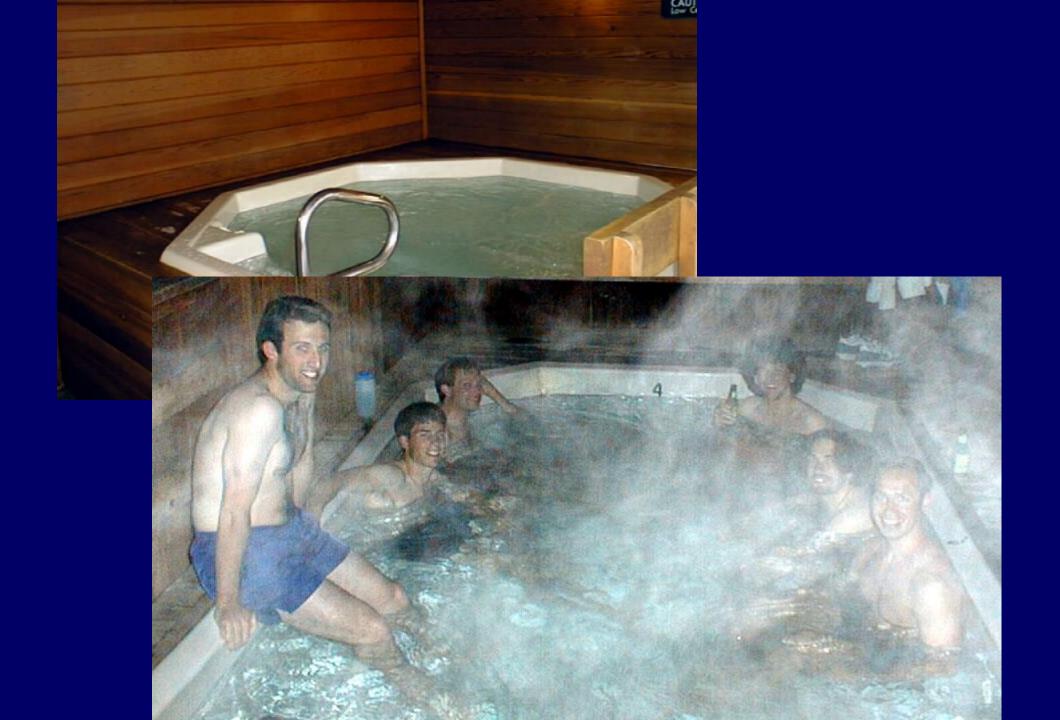




The patient was treated with corticosteroids and anti-tuberculous agents.

She recovered and is well, several years after this episode.



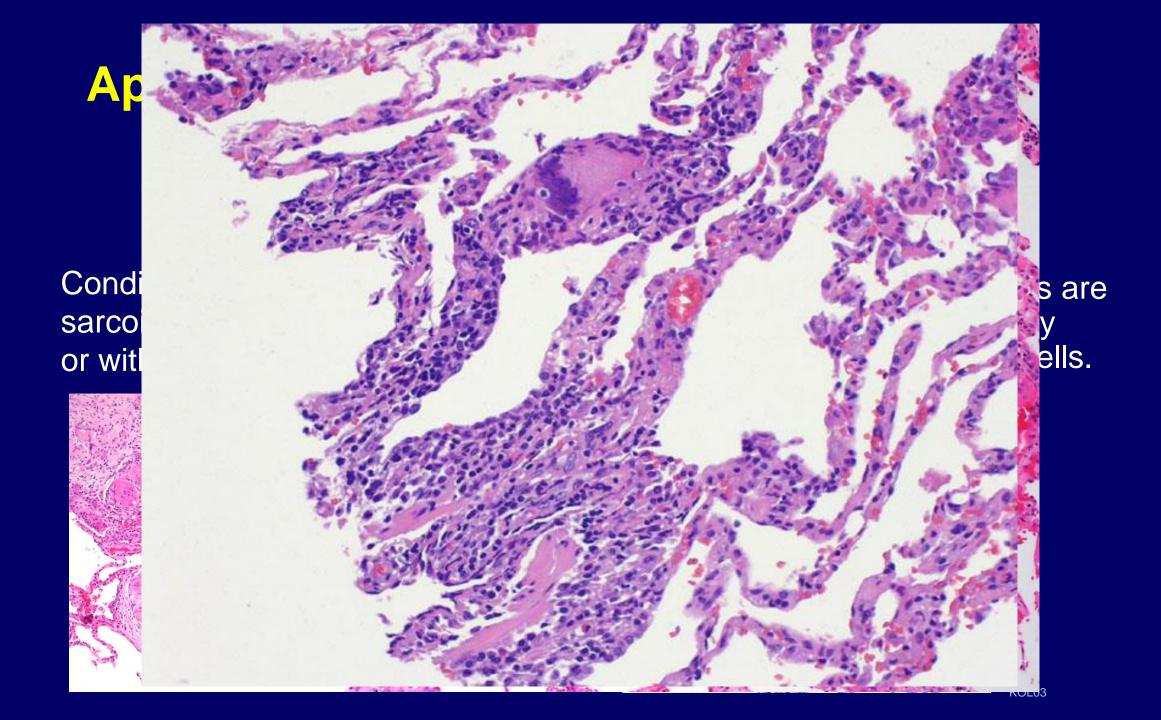


The Wide Spectrum of M. Avium-Associated Lung Disease

- 1. Fibrocaseous disease similar to MTb
- 2. Disseminated infection in the immunocompromised host
- 3. Localized/patchy nodular disease complicating bronchiectasis and other chronic lung disease.
- 4. Diffuse interstitial/nodular disease in normal hosts (so-called "hot tub lung")
- 5. Eosinophilic pneumonia (very rare)

Commentary





Simply identifying granulomas in a surgical specimen is inadequate for a final diagnosis.

It is always important to attempt to characterize:

- 1. The distribution and qualitative features of the granulomas
- 2. Any other accompanying findings and results of special stains.

Approach to granulomatous lung disease

Common diffuse granulomatous diseases:

- 1. Infections
 - Mycobacteria
 - Fungi (including pneumocystis)
- 2. Sarcoidosis
- 3. Hypersensitivity pneumonitis

4. Aspiration

One Hundred Consecutive Granulomas in a Pulmonary Pathology Consultation Practice

Julianne Roberta Hutton Klein, MD,* Henry Dale Tazelaar, MD,† Kevin Orr Leslie, MD,† and Thomas Vail Colby, MD†

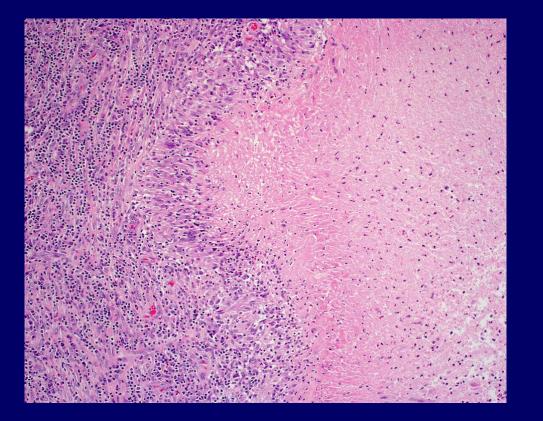
(Am J Surg Pathol 2010;34:1456–1464)

Uncommon causes of granulomatous lung disease

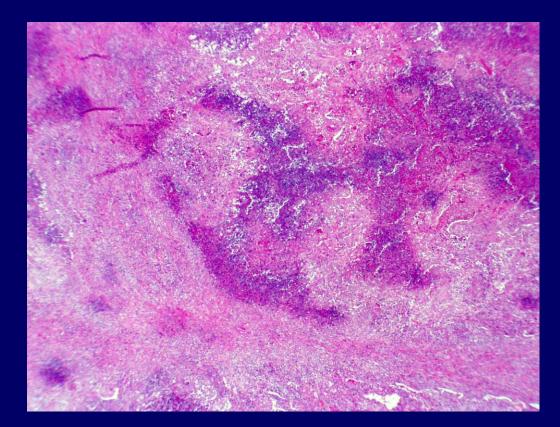
Bronchiectasis Berylliosis and other pneumoconioses **Drug reactions** Collagen vascular diseases (e.g. Sjogren syndrome) Intravenous durg abuse with crushed tablets Vasculitis (rarely) Granulomatosis with polyangiitis, Churg-Strauss syndrome Eosinophilic pneumonia Immunoglobulin deficiency (CVID) Diffuse lymphoid hyperplasia (regardless of cause) Giant cell interstitial pneumonia (hard metal disease) Diffuse neoplasms (lymphoma, leukemia, other) Inflammatory bowel disease Incidental granuloma (remote infection, local airway obstruction)

Comparison of granulomatous diseases

<u>Infection</u>



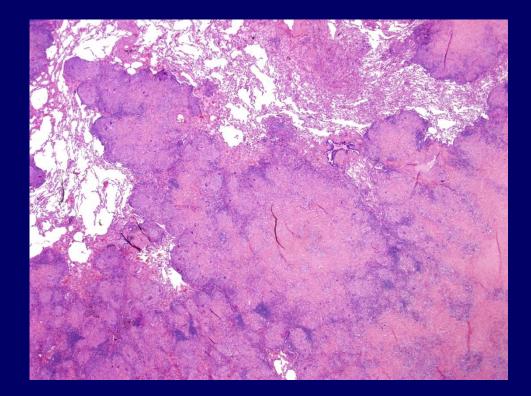
Granulomatosis with polyangiitis (GPA)

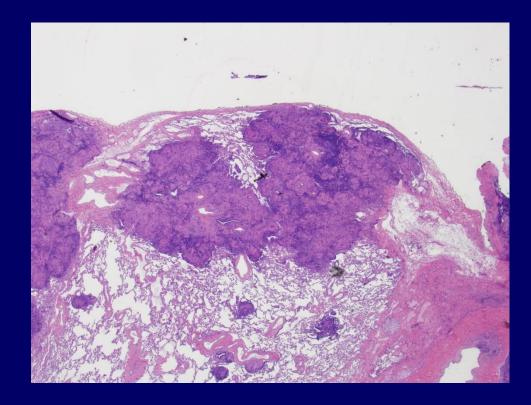


Comparison of granulomatous diseases

<u>Sarcoidosis</u>



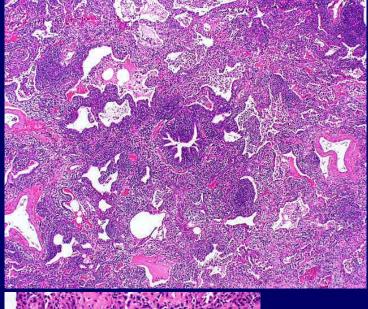


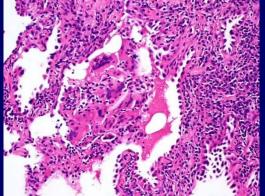


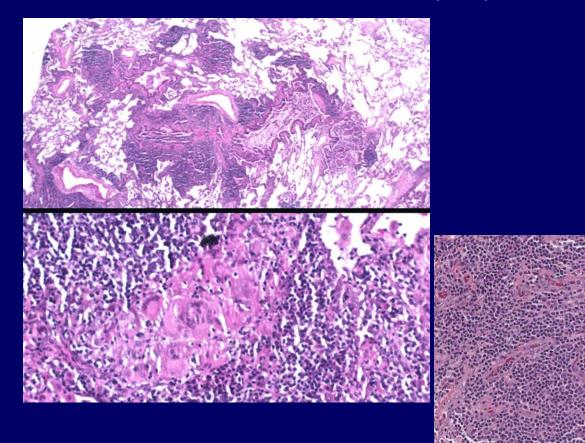
Comparison of granulomatous diseases

Sjogren Syndrome

Immunodeficiency syndromes (GLILD)







Commentary

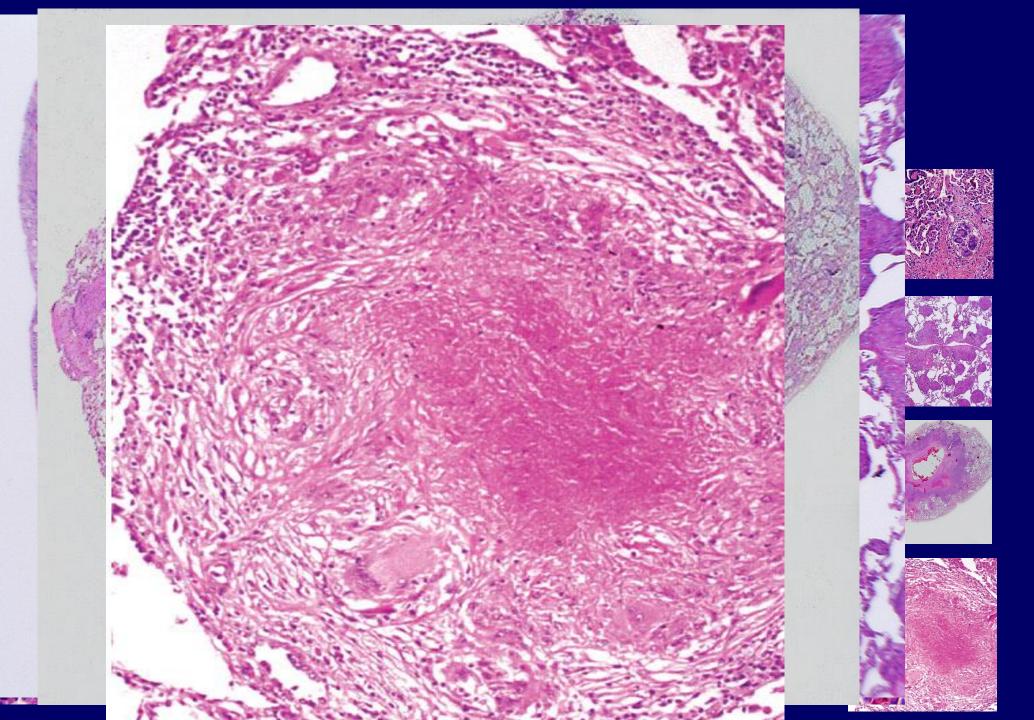


Airway-

Granulo

Angioce

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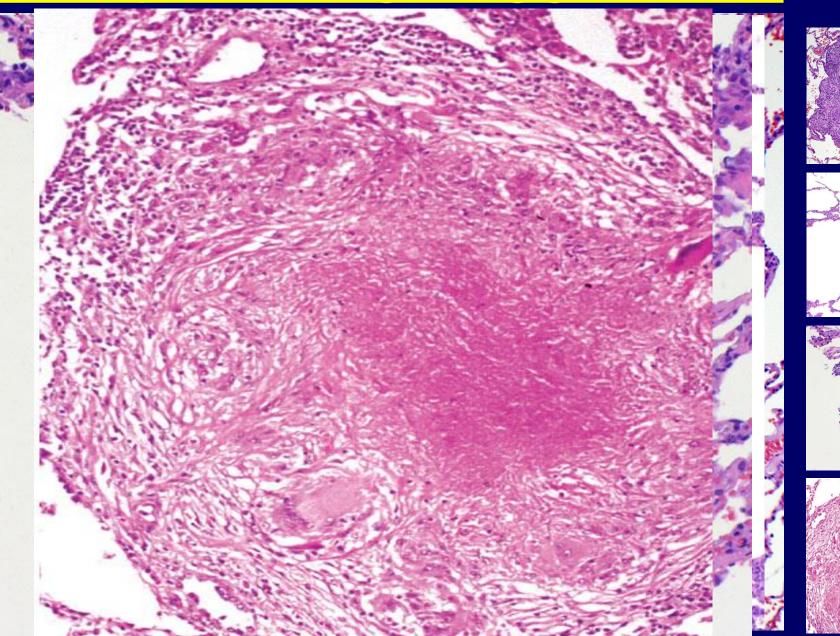
Critical questions regarding granulomas

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Critical questions regarding granulomas

Granulomas scattered and single?

Granulomas coalescent?

Birefringent material and secondary structures associated with the granulomas?

Extensive fibrous tissue around the granulomas?

Is there a mononuclear cell infiltrate associated with the granulomas?

Infection Aspiration

Sarcoidosis

nonspecific

Sarcoidosis, Chronic aspiration

Infection, Hypersensitivity Lymphoprolif.

Take Home Lessons

Recognizing granulomas in lung is child's play!

Anatomical location and appearance of granulomas provide key clues to accurate diagnosis!

Surgical Pathology of Granulomatous Interstitial Pneumonia

Oi Yee Cheung, MD, J. R. Muhm, MD, R. A. Helmers, MD, Marie-Christine Aubry, MD, Henry D. Tazelaar, MD, Andras Khoor, MD, Kevin O. Leslie, MD, and Thomas V. Colby, MD

A granulomatous interstitial pneumonia is a form of diffuse lung disease in which granulomas are a component of the histologic findings. The differential diagnosis is quite broad, but most cases represent examples of either sarcoidosis, diffuse granulomatous infections, or hypersensitivity pneumonitis. "Hot tub lung" is a recently described form of granulomatous interstitial pneumonia that appears to have some features of diffuse infections and some features of hypersensitivity pneumonitis. The pathologist's approach to these conditions can be facilitated by giving careful attention to the anatomic distribution of the granulomas, the qualitative features of the granulomas, and the histologic changes in the lung tissue around and away from the granulomas. These features, along with the results of cultures and special stains for micro-organisms and clinical and radiologic correlation allow for a diagnosis in the vast majority of cases.

Ann Diagn Pathol 7: 127-138, 2003. Copyright 2003 Elsevier Inc. All rights reserved.

Commentary



References

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Diffuse pulmonary disease caused by non-tubeculous mycobacteria in immunocompetent people (Hot tub lung).
Am J Clin Pathol 2001; 115:755-762.

Cheung OY, Muhm JR, Helmers RA, Aubry MC, Tazelaar HD, Khoor A, Leslie KO, and Colby TV: Surgical pathology of granulomatous interstitial pneumonia. Ann Diag Pathol 7(2):127-138, 2003

Hutton Klein JR, Tazelaar HD, Leslie KO, Colby TV:

One hundred consecutive granulomas in a pulmonary pathology consultation practice.

Am J Surg Pathol. 2010 Oct;34(10):1456-64.

AUDIENCE

Comments and Questions?

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