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Hypersensitivity pneumonia: UIP/IPF histopathologic presentation

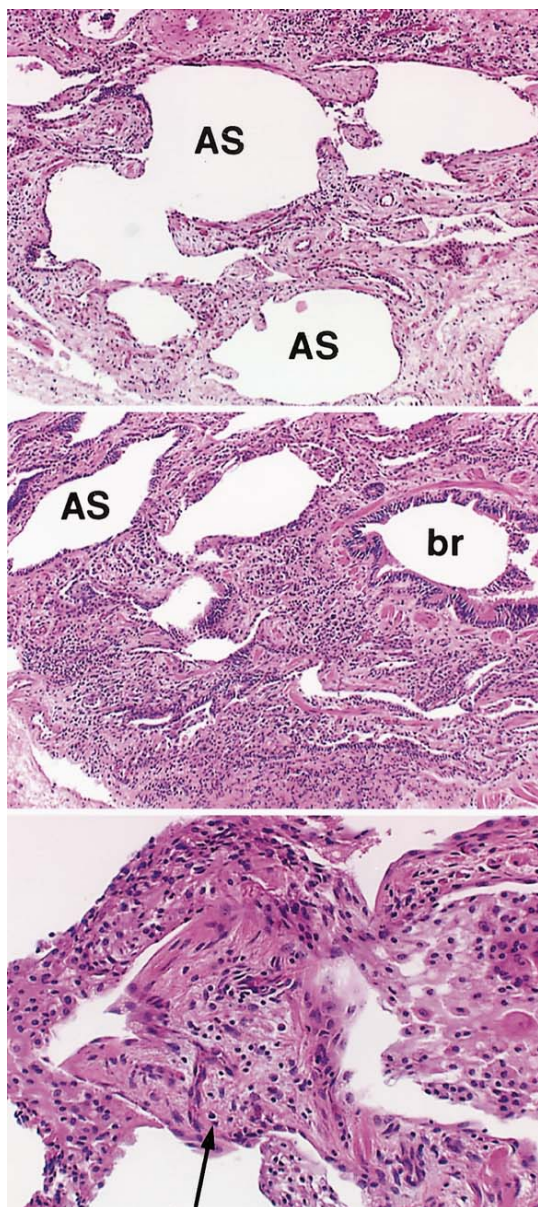
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To the editor:

A 59-year-old businesswoman had enjoyed lifelong excellent health until 7 years previously, when episodes of pneumonia had occurred in 3 consecutive years along with a chronic cough. Two years later, she had been diagnosed by means of a chest roentgenogram with interstitial lung disease, and her symptoms had progressed to include shortness of breath with activity. After worsening of pulmonary functions, she underwent an open lung biopsy.

Histopathologic assessment revealed a pattern consistent with usual interstitial pneumonia (Fig 1 ■).

Fig. 1. Open lung biopsy. **A**, The distal lung parenchyma in the subpleural zone shows early honeycombing with architectural loss of normal structures. The formation of cystic air spaces (AS) and an admixture of predominately chronic inflammatory cells and fibroproliferation in the alveolar walls are seen. **B**, In a more cellular interstitial response, abundant inflammatory cells and fibroproliferation are present. **C**, A rare focus of intramural organization is evident (*arrow*). In addition to the thickened alveolar walls, foci of alveolar macrophages and type-2 epithelial cell hyperplasia are evident. Sites of normal lung were not identified in the submitted biopsy specimen. *br*, Bronchiole. (Hematoxylin and eosin; original magnification, 40×, 40×, and 100×, respectively.)



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Treatment with systemic corticosteroids and cytotoxic agents did not seem helpful, and deterioration of volumes and diffusion accelerated. Worsening of pulmonary function test results, increased intensity of cough, and shortness of breath with minimal activity despite chronic systemic steroids led to initiation of treatment with interferon.

She was referred for an opinion concerning the possibility that workplace and/or home microbial contamination had played a causative role in this disorder. Pulmonary function testing revealed a forced vital capacity of 1.52l (49%) and a carbon monoxide diffusion in the lung of 6.2 mL/mmHg/min (33%). Abnormal laboratory data included a sedimentation rate of 42 mm/h, C-reactive protein of 1.6 mg/dL, total IgA of 609.7 mg/dL, and positive precipitins to *Aureobasidium pullulans*, *Cladosporium herbarum*, *Alternaria tenuis*, *Curvularia spicifera*, *Aspergillus niger*, *Stemphylium solani*, *Rhizopus nigricans*, *Phoma herbarum*, *Epicoccum nigrum*, *Trichoderma viride*, *Neurosporum intermedia*, and *Helminthosporium interseminatum*.

The environmental history revealed that she had worked in an old building with recurrent roof leaks that had a strong “moldy” smell. The building was condemned and she was moved to a different workplace 2 years after onset of symptoms. The home environment also had a history of recurrent roof leaks and recurrent blockage to the air conditioning condensate drainage line. An investigation of the home environment showed evidence of water damage in the ceiling in 2 areas. The condensate line was drained into an open sewage line. The plenum and proximal ducts revealed only scant mold by direct visualization. The condensate pan was found to retain approximately one eighth inch of water.

Cultures obtained through use of an Andersen sampler (Thermo Andersen Samplers Inc, Atlanta, Ga) revealed >100 total colonies/ft³ of mold and bacteria. A mycologist identified *Cladosporium* species, *Aspergillus niger*, *Penicillium* species, *Alternaria* species, and *Paecilomyces* species as most numerous. Bacteria were not identified.

The patient was moved into a proven-safe environment while remediation efforts were undertaken on her home. She remained short of breath, continued to cough, and experienced extreme tiredness and fatigue, but she continued to work full time.

Usual interstitial pneumonia (UIP), recently renamed *idiopathic pulmonary fibrosis* (IPF) in an International Consensus Statement,¹ is considered a specific form of chronic fibrosing interstitial pneumonia of unknown cause with a histologic pattern of UIP on open lung biopsy. Major and minor criteria are given to aid in the differentiation from other interstitial lung diseases of known cause that might closely mimic IPF. Chronic insidious hypersensitivity pneumonia (HP) is one such disorder that can be difficult to differentiate by clinical presentation,^{1 2} high-resolution computed tomography scans,³ and open lung biopsy.^{2 4 5} Despite the remarkable similarities of these 2 disorders, there is a striking difference in morbidity and mortality when HP is appropriately diagnosed and the causative

antigen(s) avoided. There is, however, a paucity of data in the current literature concerning environmental investigation or the assessment of antibodies directed toward environmental antigens in patients diagnosed with IPF that could prove vital in changing the diagnosis to IPF secondary to HP.

The condition of the patient described has progressed to the point at which long-term survival is unlikely despite her having moved from her contaminated home.

Although the presence of specific antibodies is not diagnostic of HP and simply reflects significant exposure, other factors that might have altered the prognosis include the following: (1) open lung biopsy earlier in the course of the disease to clearly identify the histopathologic pattern of hypersensitivity pneumonia, (2) an in-depth environmental history of both the home and the workplace since onset of symptoms, (3) an environmental investigation to identify the source and specific microbes involved, and (4) remediation of or moving away from the contamination.

All patients with idiopathic interstitial lung disease must be suspected of having a hypersensitivity pneumonia to a home or workplace contaminant until aggressive evaluation proves otherwise.⁶

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Robert L. Jacobs, MD
Biogenics Research Institute
8229 Fredericksburg Rd
San Antonio, TX 78229

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