A Curious Case of Biopsy Proven Usual Interstitial Pneumonia

Austin Makdia[1], Vishesh Persaud[1], Saiara Choudhury[1], Asad Chohan[1], Abhay Vakil[1]

1 Department of Pulmonary Medicine, HCA Corpus Christi Medical Center, Corpus Christi, Texas

CORPUS CHRISTI MEDICAL CENTER

An HCA Affiliated Hospital

Background

Usual interstitial pneumonia (UIP) refers to a combination of radiologic and histologic findings which include patchy interstitial fibrosis with alternating area of normal lung, temporal heterogeneity of fibrosis characterized by scattered fibroblastic foci with dense acellular collagen, and architectural distortion due to chronic scarring or honeycomb change [1]. Idiopathic pulmonary fibrosis (IPF) and UIP are terms that are often used interchangeably. However, UIP is not the same as IPF which requires exclusion of underlying clinical conditions including but not limited to connective tissue disorders, hypersensitivity pneumonitis, heavy metal exposure and several others. High-resolution computed tomography (HRCT) has improved diagnostic accuracy and has modified the role radiologic imaging has in managing patients.

Case Description

A 65-year-old-male patient with no significant past medical history who presented to the clinic for evaluation of a six-month non-productive intermittent chronic cough; which occurred throughout the day and night. There was no relation with any specific exposures, aggravating or relieving factors. The only associated symptom was progressively worsening shortness of breath and dyspnea on exertion; unable to walk to his mailbox without stopping. He denied any significant environmental, occupational, pet exposures and was up-to-date with immunizations. He was a lifetime non-smoker, without any alcohol or drug use. Physical exam revealed bilateral basilar crackles without any wheezing, rhonchi or cardiac, skin, joint, or eye findings.Pulmonary function testing was consistent with moderate restriction with six-minute-walk testing showing severe activity limitation with desaturation requiring oxygen.HRCT illustrated findings consistent with the indeterminate UIP pattern including bilateral interlobular and intralobular interstitial thickening, subtle reticulation with areas of ground glass opacities without any associated bronchiectasis or honeycombing (Figure 1). The patient had bronchoscopy, immunologic, and hematologic laboratory studies reported to be unrevealing. A surgical lung biopsy was performed with multiple samples collected from the right lung. Histopathologic examination showed widespread extensive destruction of the normal lung parenchyma, with largely subpleural old dense fibrosis containing numerous honeycombing areas, fibroblastic foci, without granulomas or necrosis, consistent with UIP (FIgure 2).

Case Description (continued)

The patient was diagnosed as having IPF due to negative work up and started on pirfenidone therapy, but ultimately failed and underwent bilateral lung transplantation with uncomplicated post transplant course and improved functional status.

FIgures

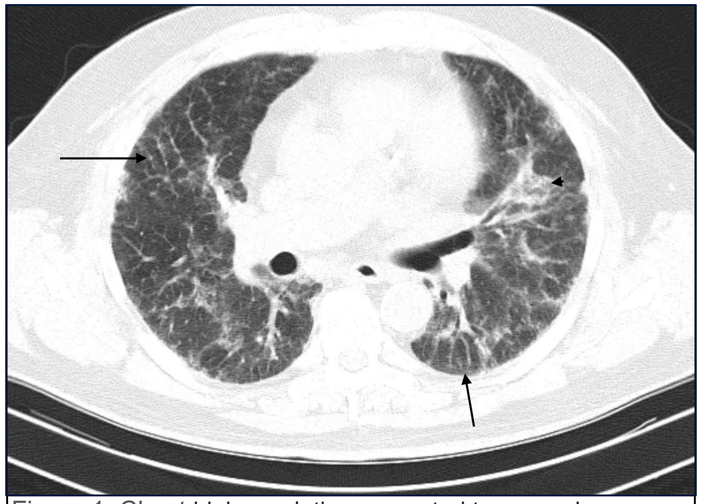

Figure 1: Chest high-resolution computed tomography showing, reticular fibrosis (arrows) also with ground glass opacities (arrow head)

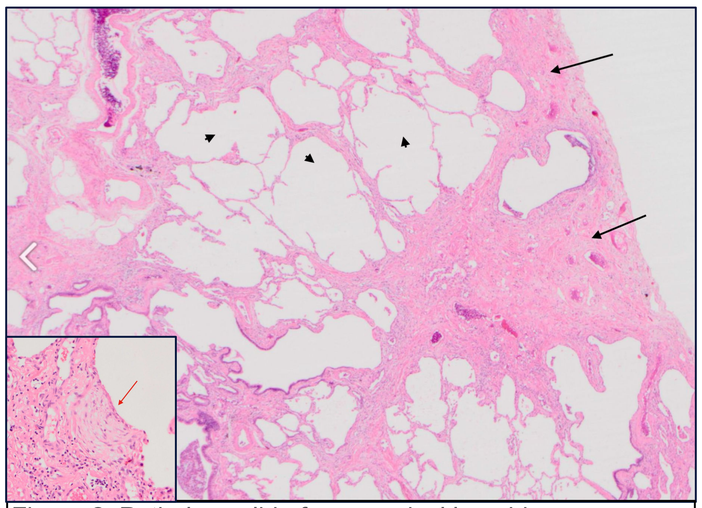

Figure 2: Pathology slide from surgical lung biopsy, subpleural dense fibrosis (arrows) also with honeycombing (arrow head), and a fibroblastic foci (red arrow)

Discussion

The new 2018 guideline categories of UIP do help eliminate the need for surgical lung biopsy (SLB), when characterizing a patient “indeterminate UIP” and “alternative diagnosis”, the clinician should be cautious about prolonging SLB in patients to determine fibrosis pattern [2]. For patients with atypical radiologic findings, the SLB with histopathologic diagnosis remains a critical step to obtain a diagnosis of UIP and is the single most important predictor of prognosis at the time of diagnosis and therefore continues to be the gold standard for diagnosis [3]. A study done by Brownell, et al. found that 22.7% of patients with inconsistent UIP radiologic findings, had definite/probable UIP on biopsy [4]. Another study by Tomassetti et al. investigated how SLB changed diagnostic confidence and changed therapeutic strategy in one-third of cases determined to be IPF or non-IPF fibrotic Interstitial lung disease (ILD) [5]. Change in the treatment and overall prognosis of patients can occur due to SLB confirming UIP pattern on histology from treatment with antifibrotics, symptomatic management, pulmonary rehabilitation and early referral to transplantation center. In our case, the radiological findings were not consistent with definitive UIP. However histopathology did establish a definitive diagnosis changing our management to initiation of antifibrotics and leading to early referral for lung transplantation.

References

1. American Thoracic Society, & European Respiratory Society (2002). American Thoracic Society/European Respiratory Society International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias. This joint statement of the American Thoracic Society (ATS), and the European Respiratory Society (ERS) was adopted by the ATS board of directors, June 2001 and by the ERS Executive Committee, June 2001. *American journal of respiratory and critical care medicine*, *165*(2), 277–304. https://doi.org/10.1164/ajrccm.165.2.ats01
2. Raghu, G., Remy-Jardin, M., Myers, J. L., Richeldi, L., Ryerson, C. J., Lederer, D. J., Behr, J., Cottin, V., Danoff, S. K., Morell, F., Flaherty, K. R., Wells, A., Martinez, F. J., Azuma, A., Bice, T. J., Bouros, D., Brown, K. K., Collard, H. R., Duggal, A., Galvin, L., … American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Society (2018). Diagnosis of Idiopathic Pulmonary Fibrosis. An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. *American journal of respiratory and critical care medicine*, *198*(5), e44–e68. https://doi.org/10.1164/rccm.201807-1255ST
3. Flaherty, K. R., Thwaite, E. L., Kazerooni, E. A., Gross, B. H., Toews, G. B., Colby, T. V., Travis, W. D., Mumford, J. A., Murray, S., Flint, A., Lynch, J. P., 3rd, & Martinez, F. J. (2003). Radiological versus histological diagnosis in UIP and NSIP: survival implications. *Thorax*, *58*(2), 143–148. https://doi.org/10.1136/thorax.58.2.143
4. Brownell, R., Moua, T., Henry, T. S., Elicker, B. M., White, D., Vittinghoff, E., Jones, K. D., Urisman, A., Aravena, C., Johannson, K. A., Golden, J. A., King, T. E., Jr, Wolters, P. J., Collard, H. R., & Ley, B. (2017). The use of pretest probability increases the value of high-resolution CT in diagnosing usual interstitial pneumonia. *Thorax*, *72*(5), 424–429. https://doi.org/10.1136/thoraxjnl-2016-209671
5. Tomassetti, S., Ravaglia, C., Puglisi, S., Ryu, J. H., Colby, T. V., Cavazza, A., Wells, A. U., Pavone, M., Vancheri, C., Lavorini, F., Matucci-Cerinic, M., Rosi, E., Luzzi, V., Gori, L., Rossi, G., Donati, L., Dubini, A., Piciucchi, S., & Poletti, V. (2022). Impact of Lung Biopsy Information on Treatment Strategy of Patients with Interstitial Lung Diseases. *Annals of the American Thoracic Society*, *19*(5), 737–745. https://doi.org/10.1513/AnnalsATS.202104-466OC

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