Hypersensitivity pneumonia-nonspecific interstitial pneumonia/fibrosis histopathologic presentation: a study in diagnosis and long-term management.

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BACKGROUND: Nonspecific interstitial pneumonia/fibrosis (NSIP) has been classified a form of idiopathic interstitial pneumonia/fibrosis. We have shown that cases of NSIP without demonstrable serum precipitins may be caused by inhalation of high levels of mold and/or bacteria in closed environments.

OBJECTIVE: We report a patient with a clinical and histopathologic diagnosis of NSIP without serum precipitins caused by a microbial contamination in her home. Her case was converted from an acute to an insidious clinical presentation by inadequate remediation. A prolonged avoidance-challenge technique demonstrated that this case of NSIP was a form of hypersensitivity pneumonia that was reversible by effective remediation.

METHODS: The patient was identified by compatible signs and symptoms, roentgenographic studies, pulmonary function tests, and a transbronchial lung biopsy. She was further evaluated with a detailed environmental history, serologic tests, and investigation of the home environment. An environmental avoidance and challenge technique was performed to confirm cause and effect and to determine that remediation had been effective.

RESULTS: Review of the biopsy showed NSIP and failed to reveal any non-caseating granuloma formation. Investigation of the home revealed a Cladosporium species contamination of the air conditioning system and Penicillium species beneath an entryway carpet. Serum precipitins to commercial antigens of common mold to the south Texas area were negative. Avoidance and challenge techniques confirmed the home as the causative environment in this case of NSIP. The patient has been free of signs and symptoms and has taken no medication for interstitial lung disease over the past 30 months.

CONCLUSIONS: Some cases of NSIP may be caused by inhalation of microbial antigen(s) in a closed environment. An environmental challenge technique was an effective method to determine the causative environment and confirm that remediation had been effective. Inadequate remediation may lead to symptomatic improvement, but may convert a patient from an acute to an insidious presenter. The environmental challenge obviates a need for specific challenges to determine specific causation. Remediation of or moving from an environmental contamination to achieve reversibility or prevent progression was the treatment of choice to avoid use of long-term immunosuppressive agents.