BRITISH JOURNAL OF HOSPITAL MEDICINE

Interstitial lung disease in common variable immunodeficiency

Sujoy Khan[1]

Author details can be found at the end of this article

Correspondence to:
Sujoy Khan;
sujoykhan@gmail.com

Sir,

The article on interstitial lung disease by Mudawi et al (https://doi.org/10.12968/hmed.2020.0556) requires inclusion of common variable immunodeficiency-associated granulomatous interstitial lung disease, which is mistakenly diagnosed as sarcoid-like lung disease, often several years before the immunodeficiency disorder (Cunningham-Rundles, 2010).

Common variable immunodeficiency is primarily an antibody deficiency disorder with variable T-cell abnormalities. Patients have recurrent infections with encapsulated bacteria as a result of extremely low levels of serum immunoglobulins (serum IgG <2 standard deviation below mean for age, low IgA/IgM levels) and have poor response to vaccines with no secondary causes of hypogammaglobulinaemia. Whole-exome sequencing has identified several genetic defects required for B-cell differentiation into antibody-producing cells and thus a truly heterogenous disorder.

Pulmonary complications include bronchiectasis (~75%) as a result of recurrent infections, and 50% of patients have various autoimmune manifestations (cytopenias, autoimmune haemolytic anaemia, enteropathy). This includes the lymphocytic infiltrative lung disease seen in 8–22% of patients, that is characterised by mixed obstructive and restrictive pathology that forebodes a poor prognosis in those with common variable immunodeficiency. Other infiltrative interstitial lung diseases include lymphocytic interstitial pneumonia (with alveolar septal thickening), follicular bronchiolitis (lymphoid follicles with germinal centres in bronchovascular tree) and organising pneumonia (Cunningham-Rundles, 2010; Dhalla et al, 2020). The histological features of granulomatous interstitial lung disease overlap between lymphocytic interstitial pneumonia, follicular bronchiolitis, non-specific interstitial pneumonia, lymphoid hyperplasia, MALT-oma (nodularity and peribronchovascular thickening) (Dhalla et al, 2020). Antibody-based tests (eg *Aspergillus*-specific IgG/IgE) are not helpful, as patients do not make antibodies.

While immunoglobulin replacement therapy at 400–600 mg/kg/month remains the primary treatment modality in common variable immunodeficiency, prophylactic antibiotics are required for most of these patients. Granulomatous interstitial lung disease variably responds to immunosuppression (as patients have other autoimmune features) with oral steroids, hydroxychloroquine, anti-tumour necrosis factor agents, rituximab or ciclosporin, including higher dose immunoglobulin replacement therapy (Cunningham-Rundles, 2010). Newer treatment modalities holds promise as underlying signalling pathways for progressive fibrosis are understood.

Author details

[1]Department of Immunology and Allergy, Hull University Teaching Hospitals NHS Trust, Queen's Centre, Castle Hill Hospital, Cottingham, UK

References

Cunningham-Rundles C. How I treat common variable immune deficiency. Blood. 2010;116(1):7–15. https://doi.org/10.1182/blood-2010-01-254417

Dhalla F, Lochlainn DJM, Chapel H, Patel SY. Histology of interstitial lung disease in common variable immune deficiency. Front Immunol. 2020;11(605187). https://doi.org/10.3389/fimmu.2020.605187

How to cite this article:
Khan S. Interstitial lung disease in common variable immunodeficiency. Br J Hosp Med. 2021. https://doi.org/10.12968/hmed.2021.0467