Journal of the Postgraduate Institute of Medicine 2020; 7(1): E107 1-4

http://doi.org/10.4038/jpgim.8252

Case Report

Interstitial pneumonia with autoimmune features (IPAF) and radiological findings suggestive of nonspecific interstitial pneumonia (NSIP) – a Case Report

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Keywords: nonspecific interstitial pneumonia, interstitial pneumonia with autoimmune features, immunosuppression

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Submitted: Sep 2019, Accepted revised version: July 2020, Published July 2020
Competing Interests: Authors have declared that no competing interests exist

Introduction

Interstitial pneumonia with autoimmune features (IPAF) is the recommended new nomenclature for a patient presenting with idiopathic interstitial pneumonia (IIP) who has clinical features of an underlying connective tissue disease (CTD) but fails to fulfil the current rheumatologic criteria for a known CTD [1]. The diagnosis is made by a combination of clinical, serological and pulmonary morphological features [2]. IIPs have different clinicopathologic entities, the commonest being nonspecific interstitial pneumonia (NSIP) which is associated with IPAF [3]. Here we reported a case of IPAF with radiological findings suggestive of NSIP.

Case Presentation

A 34-year-old, previously healthy mother of two from the Western Province of Sri Lanka was admitted with progressively worsening breathlessness of one year's duration. She complained of extreme breathlessness on walking about 500m [modified Medical Research Council (mMRC) dyspnoea Grade 4] for the previous one month. She had constitutional symptoms such as loss of weight and appetite with a nonproductive cough and pleuritic type of chest pain. She denied any haemoptysis or cardiovascular symptoms. There was no occupational or environmental exposure to dust, gas or chemicals.

She had noticed hair loss, but no malar or photosensitivity rash or oral ulceration. She had experienced bilateral, symmetrical, small and large joint pain for the last one year without swelling or early morning stiffness. There was no history suggestive of spondyloarthritis. She had no symptoms suggestive of other CTD such as systemic sclerosis, Sjogren's syndrome, polymyositis or dermatomyositis. There was no family history of autoimmune disease.

She was moderately built, pale but not icteric. There was Grade 1 clubbing of the fingers. There were no cutaneous features suggestive of a CTD. She did not have any features of respiratory

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distress. Chest examination revealed bilateral reduced breath sounds and inspiratory crackles. Her 6-minute walk test showed an arterial oxygen saturation (SpO_2) of 96% at rest and a SpO_2 of 89% after walking for 6 minutes. Interstitial lung disease was demonstrated by high-resolution computed tomography (HRCT) with radiological features suggestive of non-specific interstitial pneumonia (NSIP). Her lung function tests showed a restrictive type of lung disease with a FEV1 of 40%, a FEV1/FVC ratio of 90% and a diffusing capacity of lung for CO (DLCO) of 29%.

The patient had some features of connective tissue disorders but did not fit into any definitive diagnostic criteria for a particular connective tissue disorder. According to the Systemic Lupus Erythematosus International Collaborating Clinics (SLICC) criteria, she had only 3 positive features of SLE – non-scarring alopecia, antinuclear antibodies (ANA) and anti-double stranded DNA antibodies (Anti-dsDNA).

Figure 1: HRCT of chest showing a cellular NSIP pattern. Extensive ground glass opacification with septal thickening in bilateral lung fields. Sparing of sub pleural lung parenchyma

As she had radiological and laboratory evidence of active disease, she was started on immunosuppressive treatment with pulse therapy with intravenous (IV) cyclophosphamide 15mg/kg every other week for 3 doses and then 3 weekly for 3 doses and prednisolone 0.75mg/kg/day. The response to treatment was regularly assessed by the 6 minute walk test and lung function tests. In six months, her 6-minute walk test showed an arterial oxygen saturation (SpO_2) of 99% at rest and an SpO_2 of 95% after 6 minutes of walking, with Grade 2 mMRC dyspnoea.

Discussion

In 2015, the European Respiratory Society/American Thoracic Society introduced new nomenclature and criteria for patients with idiopathic interstitial pneumonias (IIPs) and features of underlying autoimmunity termed as interstitial pneumonia with autoimmune features (IPAF) [1]. Criteria for diagnosing IPAF include the presence of interstitial pneumonia on HRCT or lung biopsy, exclusion of other aetiologies, failure to fulfil the criteria of a defined connective tissue disease and at least one feature from at least two of these domains: clinical, serological and pulmonary morphological domains [2].

IIPs are chronic, progressive, diffuse parenchymal lung diseases that have similar clinical, radiologic and histopathologic features. Known causes of interstitial pneumonia should be excluded before diagnosis of IIP, such as occupational and environmental exposures, drug toxicity or CTD [2]. It is important to exclude an underlying aetiology from the clinical point of view because it often influences the management and prognosis.

The features of IIP on HRCT are similar to typical, non-specific interstitial pneumonia appearances such as basal predominant reticular abnormalities, peri-broncho vascular extension and subpleural sparing with ground-glass attenuation. Radiological features included in the IPAF morphological domain are NSIP, organizing pneumonia (OP), NSIP with OP and lymphoid interstitial pneumonia (LIP). Importantly, the above pattern is commonly found in connective tissue disease-associated interstitial lung disease (CTD-ILD), and its presence should prompt the clinician to suspect an underlying autoimmune process [4].

Our patient who was diagnosed with IIP had subtle clinical features suggestive of an underlying autoimmune process but did not fulfil the diagnostic criteria for a specific CTD [5,6]. The spectrum of CTDs include systemic lupus erythematosus, rheumatoid arthritis, systemic sclerosis, Sjogren's syndrome and mixed connective tissue disease. Although the above diseases share a common underlying mechanism of systemic autoimmunity and immune-mediated damage to organs, they have unique and distinguishing features from each other [1]. The patient had bilateral, symmetrical, small and large joint pain for one year, without swelling and morning stiffness, but did not fulfil the clinical criteria for rheumatoid arthritis. The other features included in the clinical domain of IPAF i.e. digital fissuring, distal digital tip ulcerations, unexplained digital oedema, palmar telangiectasia, Raynaud's phenomenon and unexplained fixed rash on the digital extensor surfaces were not found in our patient [2].

ANA, Anti-dsDNA, RF, Anti-Ro, and anti-La antibodies were positive. However, their presence alone does not establish the diagnosis of a characterized CTD. Other antibodies included the serological domain of IPAF are anti-CCP, anti-Smith, anti-ribonucleoprotein and anti-topoisomerase (Scl-70) antibodies.

The management of IIP depends on the severity of symptoms, radiological pattern, pulmonary function tests and coexisting diseases. In a patient with CTD-ILD, treating the underlying disease with systemic glucocorticoids and immunosuppressants will cure the IIP [7, 8]. There are no guidelines for the treatment for IPAF.

Conclusion

Interstitial pneumonia with autoimmune features should be suspected in any patient who presents with IIP and autoimmune features but does not fulfil the criteria of a defined CTD. Immunosuppressive therapy may be beneficial in extensive lung involvement,

Abbreviations

IPAF- Interstitial pneumonia with autoimmune features
IIP- Idiopathic interstitial pneumonia- IIP
CTD- Connective tissue disease
NSIP- Nonspecific interstitial pneumonia

HRCT- High-resolution computed tomography
ILD- Interstitial lung disease
IV-Intravenous

Acknowledgments

We would like to acknowledge the contribution of Dr. Chandimani Undugodage MBBS, MD, MRCP (UK), FRCP (Lon), Senior Lecturer & Consultant Respiratory Physician, Department of Physiology, University of Sri Jayewardenepura.

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