CASE REPORT

Obstructive Sleep Apnea, Sleep-related Hypoxemia, and Hypoventilation in a Case of Familial Interstitial Lung Disease: A Triple Whammy

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Abstract

Sleep-related breathing disorders (SRBDs) are known in patients with chronic respiratory diseases such as chronic obstructive pulmonary disease (COPD) and interstitial lung disease (ILD). Sleep-related breathing disorders are further classified into obstructive sleep apnea (OSA) disorders, central sleep apnea syndromes, sleep-related hypoventilation disorders, and sleep-related hypoxemia disorders. We describe a case of familial ILD with OSA, sleep-related hypoxemia, and sleep-related hypoventilation.

Keywords: Case report, Familial interstitial lung disease, Hypoventilation, Sleep.

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Introduction

Sleep-related breathing disorders (SRBDs) are present in 30–40% of patients with chronic respiratory diseases.[1] The obstructive sleep apnea (OSA) with chronic obstructive pulmonary disease (COPD) linkage, also known as the overlap syndrome, has been well known in the literature. Sleep-related breathing disorders are further classified into OSA disorders, central sleep apnea syndromes, sleep-related hypoventilation disorders, and sleep-related hypoxemia disorders.[2,3] Of these, OSA and hypoxemia have also been increasingly recognized with interstitial lung diseases (ILDs).[4] We describe a case of familial ILD with OSA, sleep-related hypoxemia, and hypoventilation.

Case Summary

A 57-year-old lady, with a diagnosed case of familial idiopathic pulmonary fibrosis (IPF), in view of clinical history and radiology was referred for evaluation. She also suffered from epilepsy, diabetes mellitus (DM), hypertension (HT), and metabolic syndrome. She was referred in view of sleepiness and breathlessness. She was symptomatic with a dry cough and progressive exertional breathlessness for 3 years which had progressed over last 2 months. She gave a history of ILD in all her four elder sisters of whom, three have succumbed to the disease. In addition, on inquiry she gave a history of snoring for the last 15 years with unrefreshed sleep, excessive daytime sleepiness, choking episodes, early morning headache and irritability. Her body mass index (BMI) was 36.7 kg/m^2. The Epworth sleepiness scale score was 10 suggestive of excessive daytime sleepiness. The pretest probability scores like the adjusted neck circumference was 41, STOP-BANG was 6 and BERLIN was 3. Thus, she had a high pretest probability for OSA. Her chest X-ray (CXR) showed bi-basal reticular changes (Fig. 1). Her computed tomography (CT) revealed an apicobasal gradient with interstitial thickening and honeycombing (Fig. 2) suggesting a usual interstitial pneumonia (UIP) pattern. The arterial blood gas (ABG) on room air read 7.36/44.8/60.3/23/91. Diagnostic polysomnography was done. It revealed an apnea–hypopnea index (AHI) of 13.5/hour, oxygen desaturation index (ODI) of 36.3/hour, average saturation of 79%, baseline saturation of 87%, and lowest saturation of 71%. The ABG

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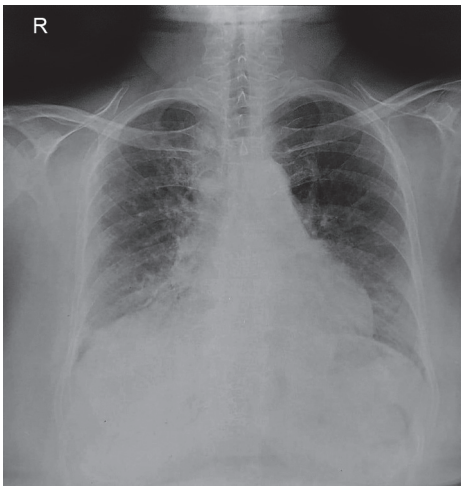

Fig. 1: Chest X-ray showing reticulonodular opacities suggestive of interstitial lung disease

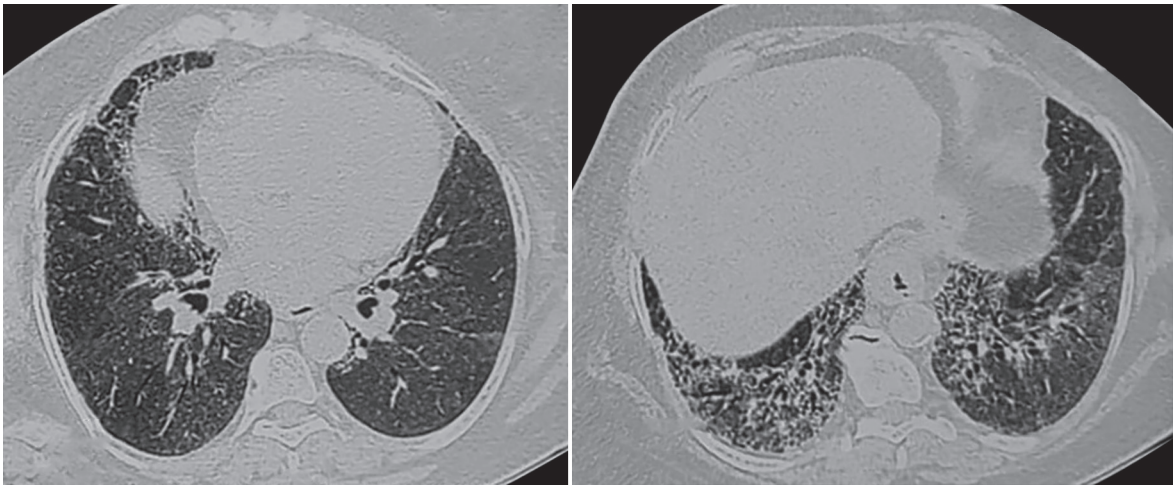

Fig. 2: High-resolution computed tomography images showing interstitial septal thickening with honeycombing suggestive of interstitial lung disease (UIP pattern)

done in sleep at night time revealed a rise in $PaCO_2$ to 55 mm Hg and read 7.32/55/58/23/89. On oxygen titration, the AHI improved to 6.3/hour, ODI to 10.7/hour, average saturation to 94%, baseline saturation to 99%, and lowest saturation to 78%. Thus, this patient was diagnosed with a case of familial ILD, and IPF with an overlap of mild OSA, sleep-related hypoxemia, and hypoventilation with metabolic syndrome, DM, and HT. She was advised nocturnal oxygen therapy.

Discussion

Interstitial lung diseases has an impact not only on the sleep architecture but also predisposes to various sleep related beathing disorders. The various forms of SRBDs reported in patients with ILD include hypoxemia, OSA, and hypoventilation. The oxygen desaturation may occur either in rapid eye movement (REM) sleep or during both non-REM (NREM) and REM sleep. Hypoxemia is an important factor in the pathogenesis of SRBD. Hypoxia-induced brainstem depression has been hypothesized to cause breathing disorders in sleep. In an awake state, ILD patients have a rapid shallow breathing pattern due to the activation of lung reflexes. Sleep causes changes in respiratory control which leads to hypoxemia. Upper airway collapsibility, and ventilatory control instability due to a multipronged pathogenesis predisposes to OSA in ILD, especially in snorers.[5–7]

Patients of ILD with nocturnal hypoxemia are candidates for nocturnal oxygen. Those with OSA are advised to use continuous positive airway pressure (CPAP). Identification and correction of sleep disturbances with supplemental oxygen during sleep has been recommended in IPF and other ILDs because this may reduce morbidity improve patient survival, and postpone the development of complications such as pulmonary HT and cor pulmonale.[5–8]

We thus describe three different types of SRBDs, that is, OSA, sleep-related hypoxemia, and sleep-related hypoventilation disorders in a single patient of ILD. In a progressive lung disease such as IPF, pulmonary rehabilitation with the management of comorbidities is the cornerstone of disease care. The impact of SRBD on ILD is multi-dimensional affecting symptomatology, disease progression, quality of life, and survival. Hence, the recognition and management of SRBD in ILD is essential to improve the quality of life and reduce mortality.

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