Case Series

Organizing Pneumonia as a Presentation of Connective Tissue Disorders

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Abstract

Organizing pneumonia (OP) is a clinical, radiological, and histological entity that is classified under the group of interstitial lung diseases. The diagnosis of OP is based on a characteristic histological pattern in the presence of certain clinical and radiological features. We present four cases of OP who presented with respiratory complaints of several weeks duration. Skin lesions typical of underlying connective tissue disorders were observed in three patients, and these were confirmed on biopsy. The diagnosis of OP was confirmed with the computed tomography scan thorax of all cases and additionally on histopathology of pulmonary lesions in two cases. All of these patients responded well to steroids, but one patient died due to respiratory failure.

Keywords: *Collagen vascular diseases, interstitial lung disease, organizing pneumonia, rheumatoid arthritis, scleroderma*

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Introduction

Organizing pneumonia (OP) is defined pathologically by the presence of granulation tissue progressing from fibrin exudates to lose collagen-containing fibroblast, in lung parenchyma and distal airways.[1] The lesions occur predominantly within the alveolar spaces but are often associated with buds of granulation tissue occupying the bronchiolar lumen (bronchiolitis obliterans). This pathological pattern is not specific for any disorder or cause but reflects progression of inflammatory process resulting from any lung injury. It may have a wide variety of causes ranging from infection, drug toxicity, lung graft reaction, radiation exposure, and connective tissue disorders (CTDs).[2] We herewith present a case series of four patients with different CTDs, viz., scleroderma, systemic lupus erythematosus (SLE), rheumatoid arthritis (RA), and Sjogren's syndrome, who presented with OP.

Case Reports

Case one

A 33-year- old male, a dye factory worker, presented with complaints of exertional dyspnea for the past 1 year that increased for the past 1 month. On clinical examination, he was breathless and had radiological features suggestive of widespread airway consolidation. His arterial blood gas analysis showed severe hypoxia. He had hard, tight skin with loss of elasticity. He also had hyperpigmented lesions [Figure 1] on abdomen and back Figure-1. A biopsy of hyperpigmented macules on the back showed histopathology features of the dermis replaced by thickened hyalinised collagen bundles extending into the subcutaneous tissue suggestive of scleroderma. His computed tomography (CT) picture [Figure 2] was typical of OP. His bronchoscopy could not be done since the patient was very hypoxic. He was started on appropriate doses of steroids and supportive treatment. Despite these and ventilatory support, the patient worsened and died of respiratory failure.

Case two

A 45-year-old female patient presented with complaints of cough and dyspnea for 6 weeks. She was diagnosed with pneumonia. In spite of repeated antibiotic courses, there was no improvement. On examination, we noticed malar rash on the face [Figure 3] and hyperpigmented discoid lesions on the back. On blood investigations, antinuclear antibody and anti-histone antibodies were positive. Her CT thorax [Figure 4] showed features

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suggestive of OP. Bronchoscopy ruled out any infective etiology. She was diagnosed with SLE with OP and started on prednisolone. The patient showed significant improvement in 10 days. Steroids were continued on tapering doses for the next 6 months.

Subsequent follow-up visits revealed complete resolution of the lung lesions.

Case three

A 45-year-old female patient presented with persistent cough and progressive dyspnea for the past 3 months. CT thorax was suggestive of bilateral air–space consolidation. Bronchoscopy and bronchoalveolar lavage (BAL) was inconclusive and did not reveal any pathogen. There was poor response to empirical antibiotics. A CT-guided lung biopsy [Figure 5] was therefore done, which confirmed diagnosis of OP [Figure 6]. On further evaluation, the patient gave a history of frequent joint pains though he did not have any obvious joint swelling or finger deformities. Her RA factor and anti-cyclic citrullinated peptide antibody were positive. A diagnosis of OP with RA was thus established. The patient responded well to corticosteroid therapy, and the lung lesions resolved on further follow-up.

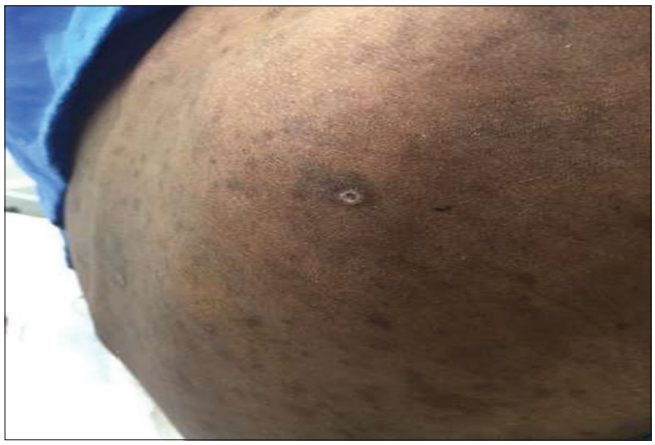

Figure 1: Skin lesions in case one

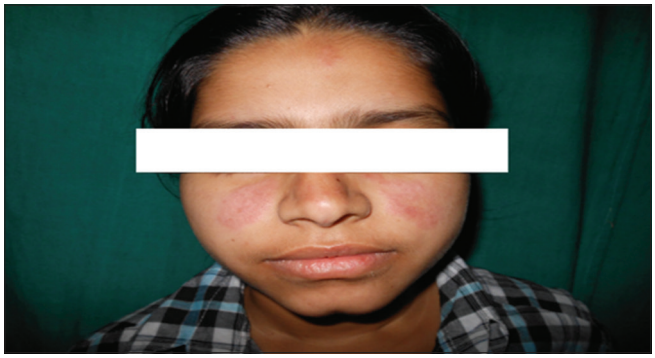

Figure 3: Malar rash

Case four

A 49-year-old male patient was referred to a case of nonresolving pneumonia. The patient had symptoms of cough and dyspnea for the past 2 months. His CT thorax showed extensive bilateral air–space consolidation with peribronchial and subpleural distribution. Bronchoscopy ruled out any significant infective cause. C-shaped arm fluoroscopy [Figure 7] guided transbronchial lung biopsy was done, which revealed alveolitis associated with fibrous plug in bronchioles [Figure 8] suggestive of organizing pneumonia. His laboratory investigations such as anti-Sjogren's syndrome–related antigen A autoantibody were found to be positive. He was put on steroids, to which he gradually responded.

Discussion

The concept of OP was introduced in the 2002 Consensus Statement of the American Thoracic Society and the European Respiratory Society for the classification of idiopathic interstitial pneumonia,[3] to distinguish cryptogenic OP from case associated with various clinical conditions related to infections, drugs, radiotherapy, and connective tissue disease.[4]

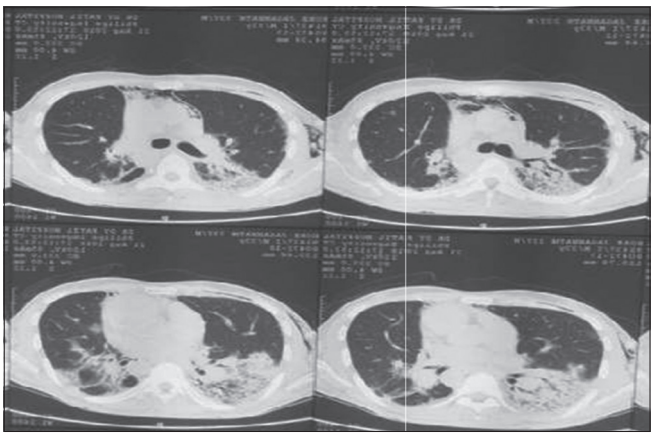

Figure 2: Computed tomography thorax images in case one

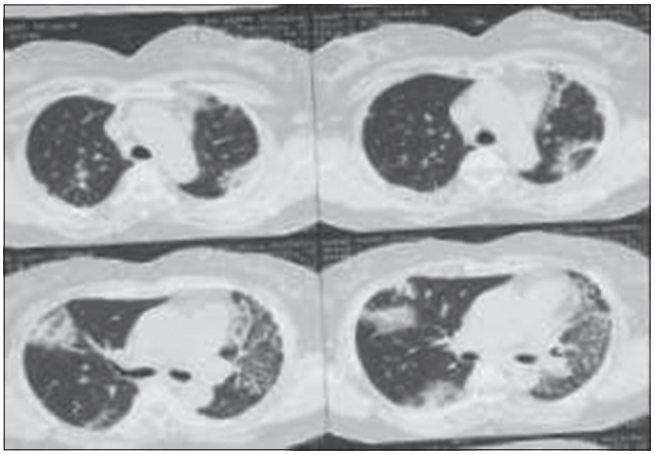

Figure 4: Computed tomography thorax images in case two

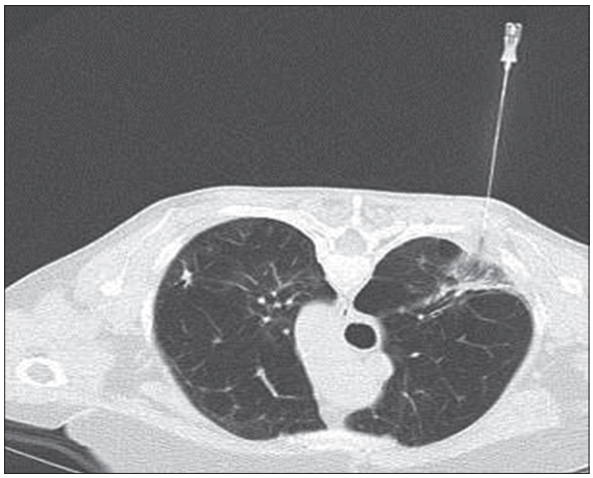

Figure 5: Computed tomography guided biopsy in case three

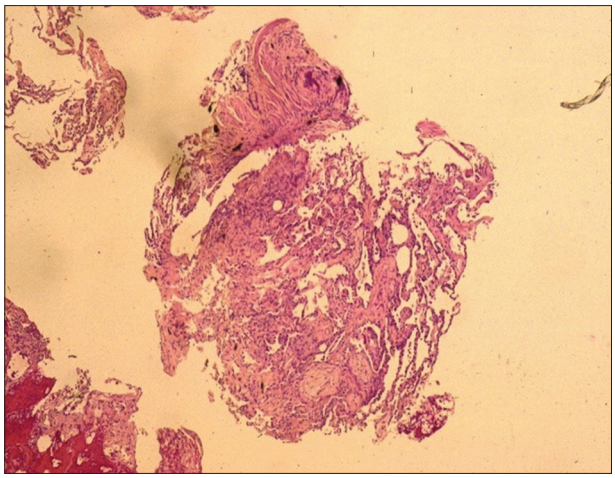

Figure 6: Histopathology picture in case three

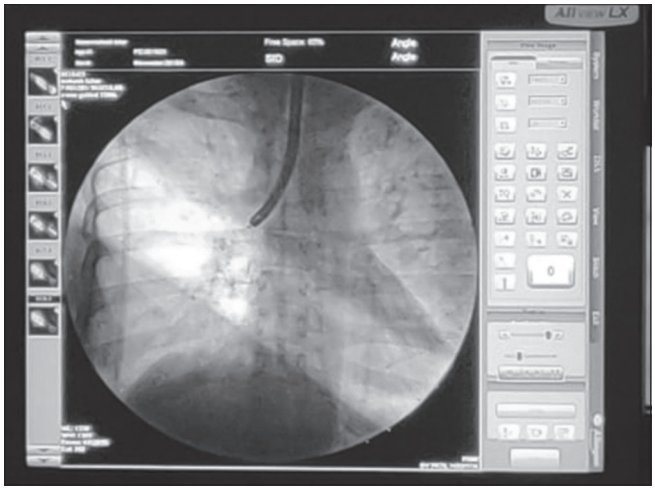

Figure 7: C-shaped arm fluoroscopy-guided transbronchial biopsy in case four

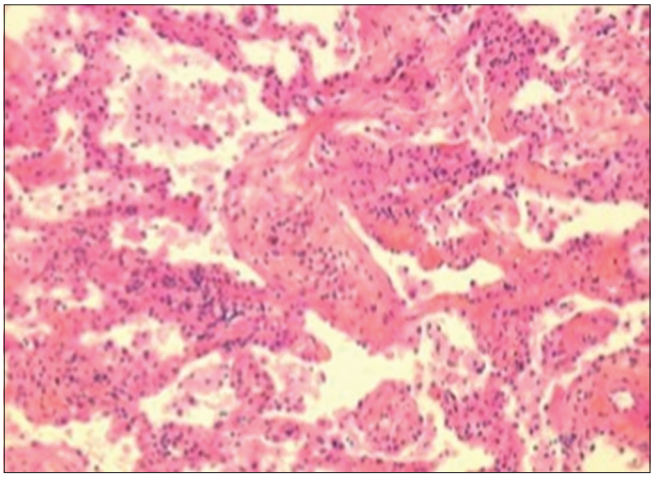

Figure 8: Histopathological picture in case four

OP is the pathological hallmark of a distinct type of lung injury and repair rather than a disease with one defined etiology.[2] Men and women are affected equally. No predisposing factors have been identified, and in particular, OP is not related to smoking. The onset of symptoms is usually subacute with fever, nonproductive cough, malaise, anorexia, and weight loss. Dyspnea is usually mild and only on exertion, but it is occasionally severe in some acute and life-threatening cases (as in our Case one). Physical examination may be normal, but fine, scattered crackles are commonly found over affected areas. The most frequent and typical imaging profile of OP is of multiple alveolar opacities with a peripheral and bilateral distribution. Bronchoscopy may be used to exclude other disorders or causes of OP, particularly infections.

The emergence of OP in the context of CTDs is considered to be a poor prognosis factor.[5] Among the connective tissue diseases, OP has been reported mostly in RA, less frequently in Sjogren's syndrome, rarely in SLE, and exceptionally in scleroderma/systemic sclerosis.[6]

All of our patients presented with dyspnea of few weeks duration, and one of them had symptoms for more than 1 year and presented with severe respiratory illness. Three patients had skin lesions typical of underlying CTDs. Bronchoscopic biopsy yielded diagnosis in one patient. All these four cases were under treatment for pneumonia initially but showed poor response. In Case one and Case two, diagnosis of OP was made on the basis of skin lesions and typical CT thorax features. However, in the third and fourth case, additional histopathological examination was required to do confirmation of diagnosis. Specific blood markers of CTDs such as RA, SLE, and scleroderma were also found positive in three patients.

Corticosteroids are the current standard treatment for OOP. British Thoracic Society guidelines[7] recommend initiating prednisone at a dose of 0.75–1 mg/kg/day. Clinical manifestations usually improve within 48 h,

but complete resolution of radiographic pulmonary infiltrates takes several weeks. Treatment of choice for OP is corticosteroids and it is entirely different from treatment of typical pneumonia. Hence, accurate and timely diagnosis are important for effective treatment in such patients.

Conclusion

OP should be considered in differential diagnosis of nonresolving pneumonia, especially in patients having features of CTDs, and should be evaluated for the same. And reverse to it, patients having diagnosis OP should be evaluated for any CTDs. Early and accurate diagnosis with proper and optimal treatment using steroids are the key for better outcome in such patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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