



BMH Medical Journal 2015;2(3):79-83 **Case Report**

Cryptogenic Organising Pneumonia As The Initial Presenting Manifestation of SLE

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Abstract:

Cryptogenic Organising Pneumonia (COP), also called idiopathic Bronchiolitis Obliterans Organising Pneumonia (BOOP), is a distinct entity among the idiopathic interstitial pneumonias defined histopathologically by intraalveolar buds of granulation tissue. The etiology includes idiopathic, infectious, drug induced radiation induced and connective tissue diseases. Organising pneumonia occurs particularly in patients with dermatomyositis-polymyositis where it may be the presenting manifestation, and rarely in SLE, rheumatoid arthritis, scleroderma and other connective tissue diseases. We describe a 30 yr old lady who initially presented with respiratory symptoms, not responding to antibiotics. She was subsequently diagnosed as SLE and HRCT thorax showed consolidation involving both lung fields. A percutaneous lung biopsy revealed features of Cryptogenic Organising Pneumonia.

Keywords: Cryptogenic Organising Pneumonia, Bronchiolitis Obliterans Organising Pneumonia, SLE

Case report

A 30-year old female was admitted in a local hospital with fever, right sided chest pain, cough and breathlessness of 5 days duration. Physical examination and chest X-ray (**Figure 1**) was suggestive of right lobar pneumonia and she was put on Coamoxyclav. In spite of antibiotics, fever was persisting and lung signs extended to left side. Repeat X-ray chest showed nonhomogenous opacity extending to the lower zones of both lung (**Figure 2**).

Hence ceftriaxone was also added to coamoxyclav. However fever was persisting and patient developed erythematous facial rash suggestive of malar rash of systemic lupus erythematosus (SLE). ANA (IIF) and antidsDNA done were found to be positive. Meanwhile the patient started developing dyspnoea and tachycardia and was referred to our centre. ECG taken was suggestive of atrial flutter. Echocardiography done showed global left ventricular dysfunction, normal valves, no vegetations. D-dimer test was negative. Patient was diagnosed as myocarditis secondary to SLE and put on ACE inhibitor and diuretics. Anticardiolipin antibody (IgG, IgM) and lupus anticoagulants were negative. Renal and liver function tests and urine examination were normal.

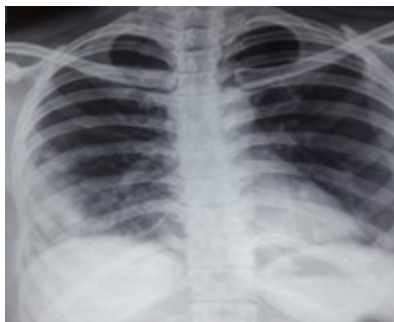


Figure 1: X ray chest suggestive of right lower lobe consolidation



Figure 2: Repeat X ray chest showing consolidation extending to left lower zone

Next day patient developed dimness of vision and optic fundus examination showed retinal vasculitic changes with haemorrhages. Patient was started on pulse methyl prednisolone (1gm IV x 3 days) followed by oral prednisolone 40mg/day along with hydroxychloroquine and azathioprine. 3 days later she became restless with violent behavior. Physical examination didn't reveal any neck stiffness or focal neurological deficits. Repeat RFT, serum electrolytes and MRI brain were normal. Patient improved with antipsychotics. Her pulmonary findings were not improving clinically. An HRCT Thorax showed segmental consolidation in the right middle lobe and both lower lobes. (Figure 3)

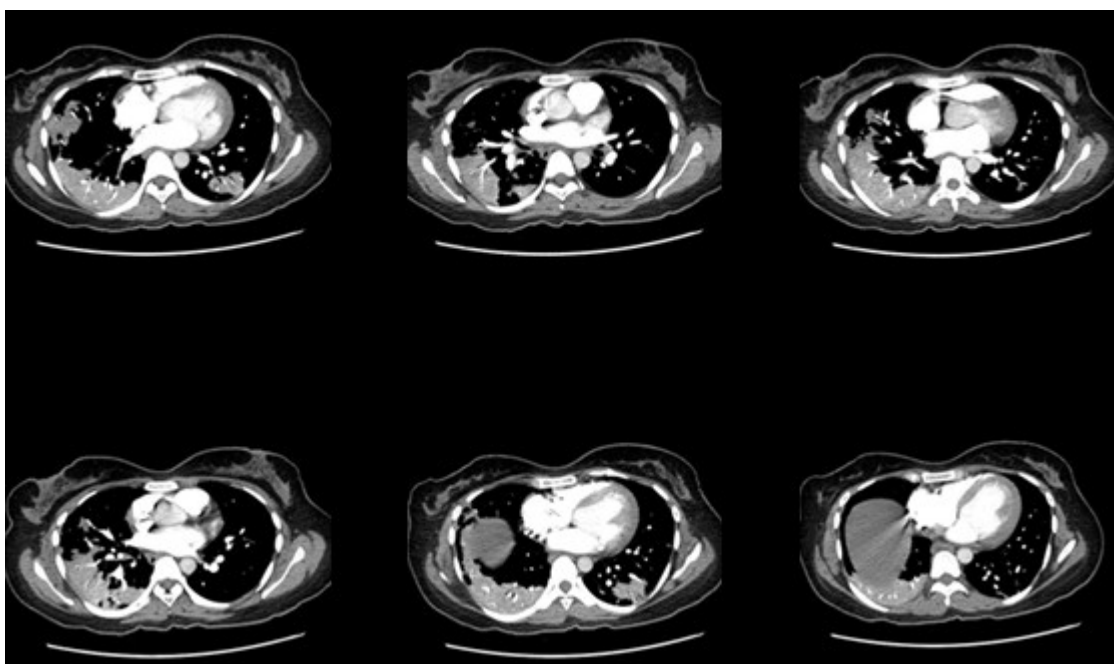


Figure 3: CT scan showing subpleural consolidation in posterior and lateral segments of right middle lobe, lateral basal segment of right lower lobe, superior and posterobasal segment of both lower lobes.

The differential diagnosis of CT Findings were lupus pneumonitis, infections, alveolar haemorrhage or organizing pneumonia. As the patient was not willing for bronchoscopy with bronchial lavage, a transthoracic lung biopsy was done. The biopsy showed alveolar air spaces filled with proliferating fibroblasts and polypoidal intraluminal fibrocollagenous plugs (Masson bodies) suggestive of Cryptogenic Organizing Pneumonia (COP). (**Figure 4**)

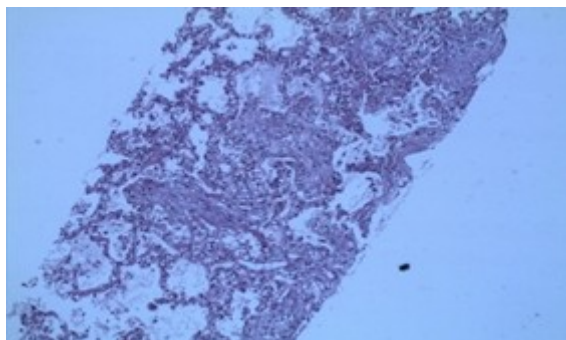


Figure 4: Lung biopsy showing alveolar airspaces filled with proliferating fibroblasts and polypoid intraluminal fibrocollagenous plugs (Masson bodies)

Patient was continued on oral prednisolone (1mg/kg/day) Hydroxy chloroquine and Azathioprine were also continued. Patient showed a slow but steady improvement. An X ray chest taken after 4 weeks showed marked clearance of the lung shadows (**Figure 5**)

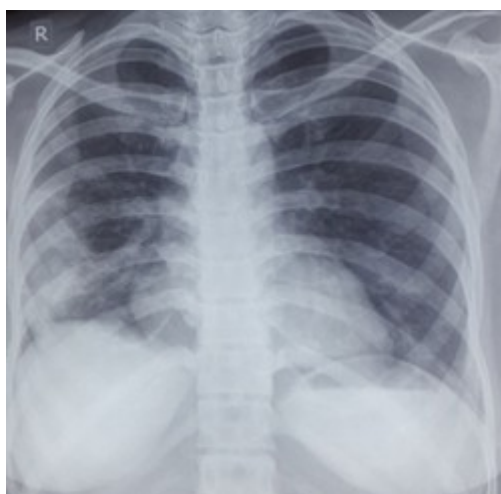


Figure 5: Xray chest one month after starting steroids showing marked clearance of lung shadows

Discussion

Cryptogenic Organising Pneumonia (COP) is classified as a distinct entity among the idiopathic interstitial pneumonias [1]. The mean age of onset is 50-60 yrs, not related to smoking. Clinically presents with fever, cough, malaise and dyspnea. Since the manifestations are nonspecific, diagnosis is often delayed. HRCT scans can be used to aid diagnosis. The main imaging patterns include multiple alveolar opacities (typical COP), solitary opacity (focal COP), and infiltrative opacities (infiltrative COP) [2].

Definitive diagnosis of COP requires a lung biopsy. The hallmark of organizing pneumonia is the presence of intraalveolar buds of granulation tissue consisting of fibroblasts-myofibroblasts embedded in connective tissue. These buds may extend from one alveolus to the next through the interalveolar pores and into the bronchioles obstructing the lumen (bronchiolitis obliterans). The

pathogenesis involves alveolar epithelial injury as the initiating event, with necrosis and sloughing of pneumocytes resulting in the denudation of the epithelial basal laminae, intraalveolar fibrinoid inflammatory cell clusters, colonization by fibroblasts and finally the formation of mature fibrotic buds, sharing many similarities with the process of cutaneous wound healing [3,4]. The etiology of COP includes infections (pneumococcal pneumonia, bacteria, viruses, parasites and fungi), iatrogenic (drug induced or radiation induced), connective tissue diseases, organ transplant, hematological malignancies, and inflammatory bowel disease.

Among the connective tissue diseases, Cryptogenic Organising Pneumonia occurs particularly in patients with dermatomyositis-polymyositis where it may be the presenting manifestation [5]. It has also been reported in rheumatoid arthritis, scleroderma, CREST syndrome and Sjogren syndrome.

Cryptogenic Organising pneumonia is rarely an initial presenting manifestation of Systemic Lupus Erythematosus [SLE], and there are only few case reports in the literature [6-9]. The pulmonary manifestations of SLE are varied and include acute lupus pneumonitis, diaphragmatic dysfunction and shrinking lung syndrome, cavitating pulmonary nodules, pulmonary hypertension, pulmonary vasculitis, pulmonary embolism, alveolar hemorrhage, chronic interstitial pneumonitis, opportunistic pulmonary infections, drug toxicity from immunosuppressive therapy and bronchiolitis obliterans (with or without organizing pneumonia) [10]. Hence the importance of a tissue diagnosis cannot be overemphasized.

Corticosteroid treatment results in slow but steady clinical improvement and clearing of the opacities on chest imaging without significant sequelae, as in our case. However relapses are common upon stopping or reduction of steroids, thus often requiring prolonged treatment.

Conclusions

Cryptogenic Organising Pneumonia as the initial manifestation of SLE is rare and requires a high degree of clinical suspicion, along with HRCT imaging and a tissue diagnosis.

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