CLINICAL CHANLLENGES IN DIAGNOSIS LUNG PARENCHYMAL ABNORMALITILES IN PATIENTS WITH RHEUMATOID ARTHRITIS

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Background

Cryptogenic organizing pneumonia (COP) is a clinical and pathologic syndrome characterized by a “Pneumonia–like” illness with excessive proliferation of granulation tissue within small airways and alveolar ducts associated with chronic inflammation in the surrounding alveoli. The radiological manifestations are quite distinctive with a pattern of bilateral diffuse but non- homogeneous, ground-glass or alveolar opacities being present in the majority of the cases. The cases have worse prognosis associated with other connective tissue disorders especially Rheumatoid Arthritis(RA). Antibiotics are not effective in treatment of this syndrome and high dose corticosteroid therapy should be used instead.

Case Report

A 54-year old male with PMHx Rheumatoid Arthritis managed on sulfasalazine for 10 years and history of 30 pack/year of smoking. Patient admitted for increasing fatigue, calf and shoulder pain, and unintentional 11 pounds weight loss over 3 months. He was feeling lethargic and weak. Patient recently failed both outpatient oral and inpatient IV antibiotics treatment for community acquired pneumonia. At the time of presentation vitals were: Temperature 98.7 F, BP of 131/70 mmHg, RR of 18/ min, Pulse 62/ min, and SpO2 94% at room air.

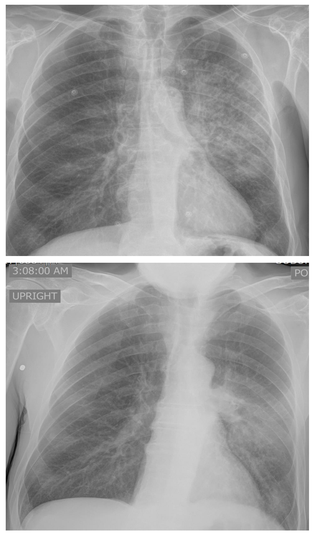
X-ray of the chest showed migratory infiltrations/ interstitial pattern of both upper and lower left lung lobes. His chest CT scan at that time was noticeable for dense consolidation in left lower lobe, several subcentimeter lung nodules bilaterally, mild left hilar and mediastinal adenopathy. Ten days prior to current admission, lung biopsy was performed to rule out malignancy. Base on the history, clinical presentation and radiology finding and lung biopsy the patient was diagnosed with COP.

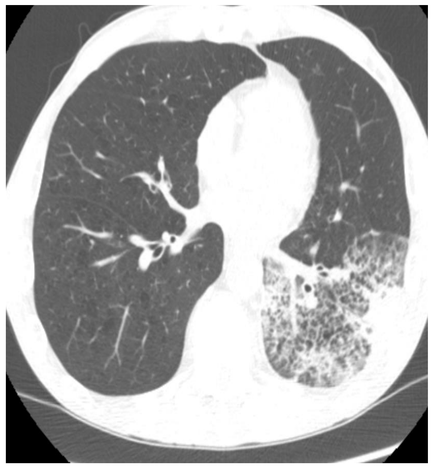
The patient was started on IV Methylprednisolone 60 mg TID for COP and free water restrictions for low Na level. His conditions rapidly improved and on the 4th post admission day, he was discharge home on 6 weeks of a tapering dose of Prednisone with instructions of smoking cessation and Rheumatology follow up.

Labs

CBC: WBC: 15.2x10*3/uL , Hb/ Hct; 11gm/32.1
Platelets; 401x10*3/uL
CMP: Na; 124 mEq/L ,K; 4.9 mEq/L ,BUN; 6mg/dl Cr; 0.5mg/dl ,CK: 31 U/L
CCP Ab IgG: >250 Units
Anti-chromatin Ab: 3.9 U and Anti-scleroderma -70 Ab:1.1 U
Lung biopsy showed marked fibrosis, with mix acute and chronic inflammation and multinucleated giant cells, and no evidence of malignancy,
Gram stains for bacteria ,acid-fast and fungal organisms were negative.

X-ray/ CT scan





References

1.Cryptogenic organizing pneumonitis. The North American experience. King TE Jr1, Mortenson RL.

2.Clinical features of organizing pneumonia associated with rheumatoid arthritis. Okada H1, Kurasawa K1,2, Yamazaki R1, Tanaka A1, Arai S1, Owada T1, Maezawa R1,2, Ishii Y1.

3.Clinical features of organizing pneumonia associated with rheumatoid arthritis. Okada H1, Kurasawa K1,2, Yamazaki R1, Tanaka A1, Arai S1, Owada T1, Maezawa R1,2, Ishii Y1.

n DJ1, Sjulin TJ2, Zingalis AA3, Skabelund AJ2.

4. Clinical Characteristics and Cytokine Profiles of Organizing Pneumonia in Patients with Rheumatoid Arthritis Treated with or without Biologics. Kawasumi H1, Gono T2, Tanaka E1, Kaneko H1, Kawaguchi Y1, Yamanaka H1.

Discussion

COP is a rare disease that occurs in both men and women with reports increasing in frequency. The typical presentation is of a subacute or chronic clinical course with fever, cough, weight loss and dyspnea, and less commonly with chest pain and hemoptysis. The diagnosis of COP requires a multidisciplinary approach combining clinical and radiological expertise, with histopathological evidence when a lung biopsy has been performed. Sometimes, COP may develop in background of connective tissue disorders especially Rheumatoid Arthritis and simulate lung cancer or coexist with lung cancer. As such, it may be difficult but necessary to make differential diagnosis between COP and lung carcinoma.

Typical radiological evidence of COP includes the presence of opacity, or consolidations with ground-glass appearance and lesions that may migrate, with bilateral and peripheral distribution. COP is characterized by granulation tissue plugs within the lumens of small airways extending into the alveolar ducts and airways. Inflammatory debris fill the alveoli and spread to the alveolar ducts and terminal bronchioles. The clusters of granulation tissue, which are composed of fibroblasts and myofibroblasts, form as a result of non-specific tissue repair.

COP is not responsive to antibiotic therapy. Treatment with systemic corticosteroids typically achieves rapid improvement in symptoms and has a >65% cure rate, without significant sequelae. The most effective therapy is long-term administration of corticosteroids at high doses for 6-8 weeks. However, when the corticosteroids are withdrawn or the dosage is decreased, COP may reoccur.

Conclusion

The patients with focal pulmonary opacity with a history of rheumatoid arthritis, smoking and immunosuppression represent a diagnostic dilemma. The presentation of COP as a solitary focal lesion is not common and occurs independently from arthritis activity at any time in RA patients. The patients with RA and abnormal CT chest findings require lung biopsy to exclude malignancy and confirm the diagnosis of COP. The most effective treatment is corticosteroids.