Dawit Kebede Huluka, Aida Mekonnen Teka, Charles B. Sherman. *Ethiop Med J, 2021, Vol. 59, No. 1*

CASE REPORT

CRYPTOGENIC ORGANIZING PNEUMONIA PRESENTING WITH ACUTE RESPIRATORY FAILURE

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Cryptogenic organizing pneumonia (COP) is one of the idiopathic interstitial lung diseases that seldom results in acute respiratory failure (ARF). We report an uncommon case of COP presenting with ARF and dramatically improving with corticosteroid treatment.

Key words: *cryptogenic organizing pneumonia, acute respiratory failure*

INTRODUCTION

Organizing pneumonia (OP) has a distinct clinicoradiologic presentation and a characteristic histologic pattern consisting of granulation tissue within the terminal or respiratory bronchioles, in the alveolar ducts, and surrounding alveoli (1). It may follow lung or systemic infections, drug toxicity, radiotherapy, autoimmune diseases, or malignancies (2). Cryptogenic organizing pneumonia (COP) is diagnosed after proper exclusion of those secondary causes of OP and is now classified as one of the acute/subacute idiopathic interstitial lung diseases by the American Thoracic Society/European Respiratory Society (3). The most common clinical presentation of COP includes cough, dyspnea, malaise, fever, weight loss, anorexia, and crackles. Patients can have consolidation, migratory infiltration, reticulonodular pattern, and mass like lesion on imaging (4). Corticosteroids remain the mainstay of treatment (4-8). We present a dramatic case of COP presenting with ARF and improving with corticosteroid administration.

CASE REPORT

A 40-year-old male farmer presented with productive cough, mild hemoptysis, dyspnea, pleuritic pain, and weight loss of two weeks duration. He had no fever, chills, skin rash, or joint pain. On admission, his PR was 122 bpm, BP 120/70 mmHg, RR 46 bpm, an O2 saturation < 70% on room air, and a temperature 36.5^{o}C. He was cyanotic and had digital clubbing and bilateral coarse crackles over the lower two-third of the chest.

His initial laboratory results were remarkable for a normal WBC; his ABG showed a pH: 7.44, PCO2: 50.1 mmHg, PO2: 37 mmHg, and HCO3 33.4 mmol/dl. Gene Xpert, ESR, HIV serology, ANA, RF, ESR, blood cultures, and cardiac ECHO were all normal. His CXR showed bilateral diffuse consolidations (Figure 1)

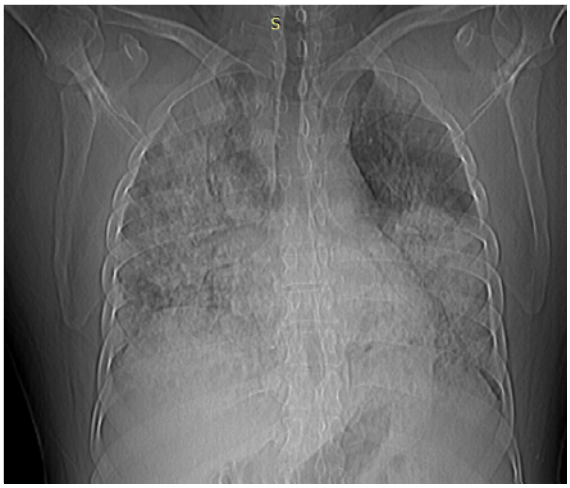

Figure 1: Admission CXR showing bilateral consolidations.

The patient was admitted to the MICU with acute respiratory failure secondary to possible ARDS; he was intubated and placed on mechanical ventilation with settings of assist control with volume control mode, FiO2 100%, TV of 6ml/kg predicted weight, RR 14 bpm, and PEEP 10mmHg. He was sedated with propofol and ketamine to RASS scale -2 to -3, started on antibiotics (Cefepime 2g intravenously three times daily and Vancomycin 1g intravenously twice daily), and begun on enteral tube feedings.

Despite all these measures, he required greater mechanical ventilator support (FiO2 100%, PEEP 16%) to maintain SaO2 89-92%. On the 4th hospital day, he had a Chest CT which revealed diffuse ground glass opacities and consolidation, and smooth thickening of the interlobular septa suggestive of COP (Figure 2). Based on these images, negative work up for connective tissue diseases, and lack of response to antibiotics, COP was considered and prednisolone 1mg/kg (60mg) po daily was added.

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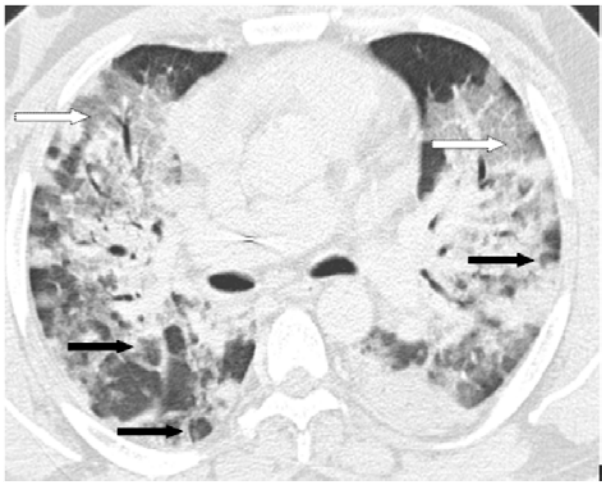

Figure 2: Chest CT showing patchy and coalescing ground glass opacity and consolidation involving all the lung lobes of the bilateral lungs, perihilar and dependent area predominance with relative sparing of the apices and anterior portions of the lung and smooth thickening of the interlobular septa (crazy paving) *(white arrows).* There are also multiple atoll (reverse halo signs bilaterally) *(black arrows).*

Over the next 5 days, he clinically improved and was extubated on the 9th hospital day. A repeat Chest CT, on 21st day of ICU admission revealed significant radiographic improvement (Figure 3). He was subsequently discharged home on prednisolone 40mg po daily, which was tapered over six months, and supplemental oxygen, which was discontinued on the tenth month.

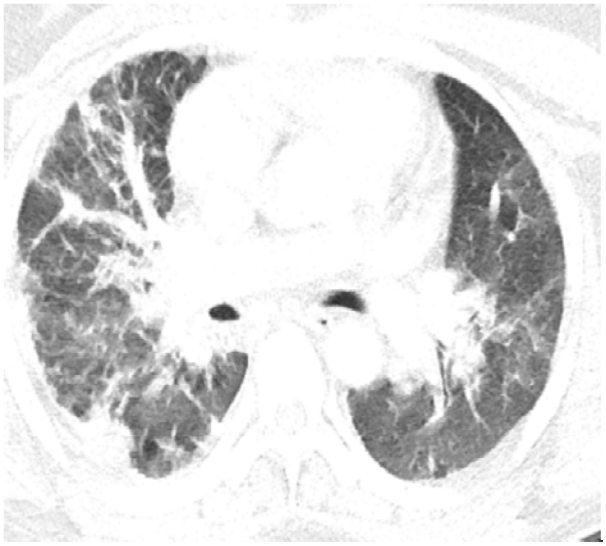

Figure 3: Repeat Chest CT three weeks later showed marked improvement in the bilateral consolidations and the perivascular and peripheral opacities.

DISCUSSION

To our knowledge, ARF due to COP requiring intubation and mechanical ventilation is an uncommon presentation (9 - 12). Although we did not have tissue confirmation (open lung biopsy is rarely done in Ethiopia) his clinical presentation, characteristic Chest CT findings, and response to steroids all make COP the most likely diagnosis. Furthermore, secondary causes of COP including infection, drugs, connective tissue diseases and malignancies, and other interstitial lung diseases like hypersensitivity pneumonitis were not supported by the laboratories or imaging. Others have also managed COP without tissue confirmation (13).

Whereas cough, dyspnea, malaise and fever are the most common manifestations of COP, weight loss, hemoptysis and chest pain are less often noted (4, 6, 14-15). Crackles are found in most cases (4, 13-15), whereas digital clubbing, as seen in our patient, is rare (4, 16). The classic radiographic form of COP is characterized by patchy, peripheral and peribronchial multifocal parenchymal consolidations (2). The atoll sign (or reverse halo sign), which denotes the presence of an area of ground glass opacity surrounded by a ring-/crescent-shaped consolidation was once considered pathognomonic of OP but is only occasionally seen (2). Crazy paving pattern is an uncommon presentation of COP and describes areas of ground-glass opacities superimposed on focal septal thickening of pulmonary parenchyma (2).

Despite the severity of illness, our patient responded successfully to treatment with corticosteroids, returning to his previous lifestyle without evidence of recurrence over two years.

Conclusion: COP can present as ARF and should be suspected in those patients with typical clinical and radiologic findings and no evidence of secondary causes.

ACKNOWLEDGEMENTS

The authors would like to thank the East African Training Initiative (eatiethiopia.org) and its sponsors, Vital Strategies New York, New York, USA and the Swiss Lung Foundation, Zurich, Switzerland, for guidance in case selection and manuscript preparation.

Conflict of interest

Each of the contributing authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

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