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Organizing pneumonia in the background of acute lymphoblastic leukemia that was successfully managed by treatment against pneumocystis jiroveci pneumonia

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SUMMARY

Organizing Pneumonia (OP) is a histopathological condition characterized pathologically by the inflammatory reactions in the bronchoalveolar wall and lumen with a multidimensional etiological feature. This phenomenon may be appeared as idiopathic or secondary to different pathological conditions such as toxic, immunologic, inflammatory, drug-related, in combination with some infectious pulmonary problems (such as pneumocystis jiroveci pneumonia or PJP), even malignant conditions. Identifying the etiological origin of OP for appropriately management is very important especially in the background of infection sources. Herein, we described a rare case of OP in the background of Acute Lymphoblastic Leukemia (ALL) that was finally managed by the therapeutic approach for PJP. We in fact faced with a case of OP in the background of ALL that was successfully managed and improved by anti-PJP treatment protocol.

Key words: organizing pneumonia, pneumocystis jiroveci pneumonia, acute lymphoblastic leukaemia

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INTRODUCTION

Organizing Pneumonia (OP) is a relatively uncommon fibrotic pulmonary disorder which characterized pathologically by the inflammatory reactions in the bronchoalveolar wall and lumen and peribronchiolar scarring, obliterating the airway lumen by polyps due to loosing connective tissue as well as accumulation and infiltration of fibroblasts and inflammatory cells particularly in bronchioles and the alveolar ducts [1,2]. This phenomenon may be appeared as idiopathic or secondary to different pathological conditions such as toxic, immunologic, inflammatory, drug-related, in combination with some infectious pulmonary problems (such as Pneumocystis Jiroveci Pneumonia or PJP), even malignant conditions [3-5]. In recent years, the traces of OP have been found following hematologic malignancies such as leukemia as well as hematopoietic stem cell transplantation [6, 7]. Herein, we described a rare case of OP in the background of Acute Lymphoblastic Leukemia (ALL) that was finally managed by the therapeutic approach for PJP.

CASE PRESENTATION

The case described was a 1-year boy suffering ALL and planned for treatment by chemotherapy based on the BFM2009 protocol in initial induction phase. Within the treatment period, the patient was hospitalized because of the appearance of fever and neutropenia followed by cough. The patient's symptoms gradually worsened as rapid and superficial respirations with decrease in arterial oxygen saturation. In chest radiological assessment (Figure 1), bilateral diffuse interstitial shadowing along with patchy alveolar infiltrates, and bronchial thickening were found. According to this appearance, chest Computed Tomography (CT) scanning was requested that disclosed the presence of multiple nodular lesions in different lobes associated with alveolar infiltrates with the diagnosis in favour of OP (Figure 2).

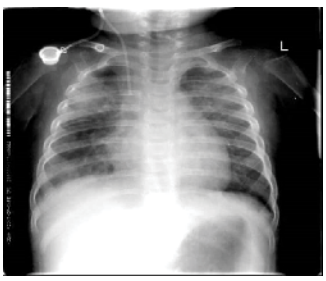

Fig. 1. There are patchy consolidation of Right lung and ground glass opacity of Left lung upper zone

The patient was thus referred for bronchoscopy and BAL fluid analysis that revealed infiltration of inflammatory cells including, alveolar macrophages, lymphocytes, eosinophils, granulocytes, as well as some contaminating erythrocytes and epithelial cells which proposed diagnosis of PJP (explaining that the evidences found on chest radiography and CT did not match the PJP diagnosis). Overall, the patient was planned to treat with parenteral co-trimoxazole plus inhaler budesonide as a choice treatment for PJP. With this treatment approach, the patient's symptoms gradually improved and completely disappeared. In final, we in fact faced with a case of OP in the background of ALL that was successfully managed and improved by anti-PJP treatment protocol.

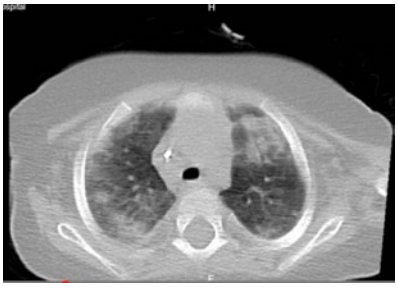

Fig. 2. There are patchy consolidation with predominantly subpleural and peribronchial distribution in both lungs (Some of these areas show reverse halo sign. These findings are suggestive of OP)

DISCUSSION

Organizing Pneumonia (OP) is a histological pattern of alveolar inflammation could be idiopathic (called COP) or secondary to infectious and non-infectious causes. Although the diagnosis of OP is established by biopsy and histology, the clinical findings and imaging changes can suggest the diagnosis. In this context, HRCT of chest is the imaging method of choice of diagnosis OP.

The most common HRCT finding of OP is patchy Ground Glass Opacity (GGO) and consolidation with a predominantly subpleural and /or peribronchovascular distribution. On the other hand, HRCT yields a high sensitivity for PJP diagnosis in immunocompromised patients.

Several studies [8-13] have evaluated HRCT findings of non-HIV PJP patients and show that they mainly exhibit ground glass opacities with several patterns of distributions include central distribution with relative peripheral sparing, mosaic pattern and a diffuse and nearly homogeneous distribution. With more advanced disease, septal line superimposed on G.O.O (crazy paving appearance) and consolidation may develop.

In our case, the chest CT scan showed bilateral consolidation and G.O.O with a predominantly subpleural and peribronchovascular distribution that is suggestive of OP.

As explained above, parenchymal opacity of PJP has not predominantly subpleural distribution and this feature is neither typical nor atypical finding of PJP. Actually, to the best of our knowledge, PJP is not a radiological differential diagnosis of predominantly peripherally located opacities.

Therefore, we believe that, regarding to CT scan and BAL findings, this case is and unusual case of OP associated with PJP. The main point of the present study was to treat OP in a case suffering ALL by using first line treatment protocols for PJP. In other words, although we did not expected to achieve complete improvement of OP following the use of parenteral co-trimoxazole, but this phenomenon was successfully managed by this treatment. Overall, both findings of OP in the background of ALL and also acceptable response rate of OP to PJP treatments are very rare. Overall, OP is a histopathological condition which can be associated with neurological, immunological, and even infectious conditions and therefore it is known as a disease with multidimensional etiological feature. Histologically, OP is mainly characterized by obliteration of the bronchiolar and alveolar lumen by organizing connective tissue [14]. Clinical conditions of the disease widely varied according to its baseline etiology as well as the disease staging. However, the prominent clinical manifestations include fever, dry cough, malaise, and dyspnea that might be gradually deteriorated. The appearance of patchy lung infiltration can be the first radiological finding. The pulmonary function may be impaired with restrictive feature. BAL also indicates an accumulation of inflammatory cells [14]. The definitive diagnosis can be achieved by open or transbronchial biopsy. The main therapeutic approach for OP is corticosteroids therapy leading complete resolution in most cases however some cases may face with treatment failure as well as high relapse rate[14]. This failure rate has been shown to be related to infectious sources such as in those with PJP needing antibiotic therapy.

As shown in our study, the patient described was definitively diagnosed as OP in the background of ALL. First, we should point that some cases have previously described cases with OP in the background of leukemia or even following organ transplantation. In a case presented by Battistini et al [15], the occurrence of OP was described in three children with ALL or promyelocytic leukemia that appeared following the treatment protocols including cytosine arabinoside and anthracyclines. In a case described by Kanda et al [16], a case with OP after syngeneic bone marrow transplantation for ALL was described that was finally managed by corticosteroid therapy. Second, as previously pointed, the main source of OP in any background (such as ALL) may be due to infectious conditions such as PJP that can be successfully managed by specific antibiotics. As shown by Kleindienst et al [17], a case of OP associated with PJP after liver transplantation and tacrolimus based immunosuppression was described that managed by antibiotic treatment. In our case, it seems that the successful management of OP using parenteral co-trimoxazole plus inhaler budesonide as a choice treatment for PJP could be related to occurring PJP in background of OP in ALL condition. In total, this phenomenon is very rare; this patient appears to be the second case of OP following PJP infection in the world and it brings us to the point that identifying the etiological origin of OP for its appropriately management is strongly recommended.

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None

CONFLICT OF INTEREST

None

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