Ramin Sami[1], Mohammadali Zohal[2], Forozan Mohammadi[3]

[1]Department of Internal Medicine, Isfahan University of Medical Sciences, Isfahan, Iran, Isfahan, Iran
[2]Pulmonary Diseases, Metabolic Diseases Research Center, Qazvin University of Medical Sciences, Qazvin, Iran, Metabolic Diseases Research Center, Velayat Hospital, 22 Bahman Blvd., Elahiyeh, Qazvin, Iran, Qazvin, Iran
[3]Department of Pathology, National Research Institute of Tuberculosis and Lung Disease, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Adult onset still disease associated with endogenous lipoid pneumonia

The authors declare no financial disclosure

Abstract

Cholesterol pneumonia or endogenous lipid pneumonia (ELP) is a rare disease that can occur in the context of a systemic disease or following a bronchial obstruction. It is characterized by a wide range of diverse symptoms and various disease course. The present report introduces a young woman diagnosed with adult onset still disease three years ago, who has been referred with macrophage activation syndrome (MAS). She underwent biopsy due to dyspnea and a crazy paving pattern in HRCT of the lungs, leading to the diagnosis of lipoid pneumonia based on the interstitial lymphocytic inflammation and cholesterol granulomas. So far, there has been no report indicating MAS associated with cholesterol pneumonia. This is the second case reporting ELP in the adult onset still disease.

Key words: endogenous lipoid pneumonia; cholesterol pneumonia; adult onset still disease

Adv Respir Med. 2019; 87: 50–53

Introduction

Lipoid pneumonia or cholesterol pneumonia is caused by the accumulation of cholesterol in the lungs. Fat could enter the lungs exogenously through the mouth and nose, which is also called *exogenous lipoid pneumonia*, or could be accumulated endogenously in the lungs [1]. It seems that the inability of macrophages to gather cholesterol collected in the alveoli due to the destruction of the type-II pneumocytes results in the endogenous accumulation of fat in the lungs [2]. The accumulation of fat in the lung tissue causes fibroblastic inflammation of the interstitial lung tissue, which can lead to progressive dyspnea and, in some cases, respiratory failure. Notwithstanding the common exogenous lipoid pneumonia, the endogenous type of this disease is rare, so that only a few cases have been reported thus far.

Adult onset still disease (AOSD) is characterized by the joints' involvement associated with systemic symptoms, which is commonly found in adolescence. Although pulmonary involvement is not frequent in AOSD, pulmonary hypertension and pleuritis are known as the most common pulmonary involvements associated with this disease.

In addition, a few cases of interstitial lung tissue involvement have been reported among these patients [3]. In this report, the described case is a patient with AOSD who developed cholesterol pneumonia.

Case report

The patient was a 22-year-old, non-smoking woman, who had been receiving prednisolone and methotrexate treatments as soon as being

Address for correspondence: Mohammadali Zohal, Pulmonary Diseases, Metabolic Diseases Research Center, Qazvin University of Medical Sciences, Qazvin, Iran, Metabolic Diseases Research Center, Velayat Hospital, 22 Bahman Blvd., Elahiyeh, Qazvin, Iran., Qazvin, Iran, e-mail: Zohal11@yahoo.com
DOI: 10.5603/ARM.a2019.0008
Received: 2.09.2018

ISSN 2451–4934