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Case Report

Interstitial Lung Disease in a 10-year-Old Child: A Case Report

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Abstract

Interstitial lung disease is rare diseases in children. Respiratory symptoms appear progressively, are often subtle and diagnosis is often delayed by many months after onset. The histological diagnosis is confirmatory but high-resolution chest computed tomography is the most sensitive imaging technique for demonstrating and identifying interstitial lung disease. Interstitial lung disease in children mostly is idiopathic though it may be caused by several conditions.

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Introduction

Pediatric interstitial lung disease is a heterogeneous group of rare diseases. It is diffuse lung diseases with chronic respiratory symptoms with a negative impact on growth.[1] It is less frequent in children than in adults and differs considerably in its etiology and pathogenesis.[2, 3] For example, desquamated interstitial pneumonia (DIP) occurs in both age groups but has different pathophysiology. It is related to prolonged smoking in adults, whereas in children it can be idiopathic, but is mostly related to an inborn error of surfactant metabolism.[4]

We are reporting a case of interstitial lung disease in a 10-year-old child who was initially treated as a patient with tuberculosis. So far, this is the first case report of interstitial lung diseases in children admitted in RMCH.

Case presentation:

A 10-year-old boy was admitted to our department of pediatrics with complaints of recurrent respiratory distress and progressive dyspnea for 7 years. He developed cough which was persistent in nature, nonproductive, aggravated after physical exertion. He developed low grade, intermittent irregular fever. He was diagnosed and treated for tuberculosis with anti-TB drugs for 6 months with no improvement. He had no family history of such types of diseases. A full work-up was done to exclude cystic fibrosis. Her physical examination revealed a respiratory rate of 50/min, a transcutaneous oxygen saturation of 92% on 1.5 L/min of supplemental oxygen by nasal cannula, chest retractions and fine inspiratory crackles at both lung field with vesicular breath sounds in both lung field. Cardiac examination was normal and there was no clubbing. Chest radiographs consistently showed multiple bilateral confluents infiltrates with air-bronchograms and a normal

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heart size. In the absence of pulmonary hypertension and cardiac malformations, chronic pulmonary disease was considered. Pulmonary function studies showed a restrictive defect. HRCT of chest revealed homogeneous ground glass attenuation with reticulation and honeycombing in the both lungs, more marked in the upper, lower and lingual lobe (Figure 1).

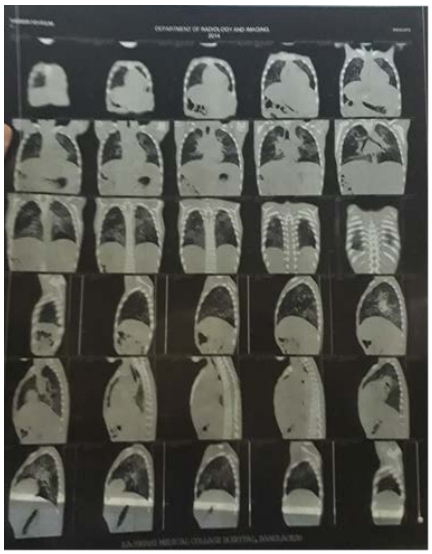

Fig-1: HRCT of chest showing homogeneous ground glass attenuation with reticulation and honeycombing in the both lung

Discussion

Idiopathic interstitial pneumonias are a group of diffuse infiltrative lung diseases of unknown etiology. It composed of several clinicopathologic subtypes including IPF also known as usual interstitial pneumonia (UIP), which is the most common subtype; nonspecific interstitial pneumonia (NSIP); respiratory bronchiolitis-associated interstitial lung disease (RB-ILD); desquamative interstitial pneumonia (DIP); cryptogenic organizing pneumonia (COP); acute interstitial pneumonia (AIP); and lymphocytic interstitial pneumonia (LIP).[5] Familial IPF is defined as when two or more members of the same family show clinical features of idiopathic interstitial pneumonia.[1,2.] The condition was first described in 1907 by Sandoz.[6] Another report[7] described IPF in identical twin sisters and since that time 15 definite cases and 3 other probable cases of pulmonary fibrosis have been diagnosed in the same family. The exact prevalence of familial IPF is unknown, however two recent studies suggest that 0.5-3.7% of all patients with IPF have disease of the familial type. The mode of inheritance of familial IPF is not entirely clear; however, the observed father-to-son transmission excludes X-chromosome linkage, whereas a single report suggested that inheritance is an autosomal recessive trait.[11] The majority of reported cases demonstrate an autosomal dominant characteristic with reduced penetrance.[1,2,8,10,12] Sporadic IPF usually presents between 50 and 70 years of age. Progressive dyspnea upon exertion, nonproductive cough, clubbing of the digits, and bibasilar end-inspiratory crackles constitute the typical clinical presentation. In a retrospective study, Lee and colleagues examined 27 patients from 15 families with familial IPF.[2] The clinical findings, pulmonary function testing, pathologic changes, radiographic features, and survival data were similar in patients with familial IPF and non-familial IPF. HRCT of chest were performed on 66% of patients and bilateral irregular linear opacities were seen in all cases. Other findings included a subpleural predominance of linear opacities (94%), subpleural traction bronchiectasis (82%), emphysematous changes (18%), ground glass opacities (12%), and bronchial wall thickening with irregularities (6%). Although the cited investigators concluded that the radiographic features of familial IPF and non-familial IPF were similar, a lower incidence of honeycombing (29%) was seen in familial IPF patients compared to those with non-familial IPF. Nishiyama and colleagues[13v]described HRCT findings in nine patients with biopsy-proven familial IPF, and found a resemblance between familial IPF and non-familial IPF, except that the prevalence of honeycombing (33%) and lower lung zone distribution (67%) was lower in familial IPF patients compared to those with non-familial IPF. In this case HRCT of chest revealed homogeneous ground glass attenuation with reticulation and honeycombing are noted in the both lungs more marked in the upper, lower and lingual lobe. Age at onset appears to be lower in patients with familial IPF, who usually present with symptoms

during the third or fourth decade of life.[14] This probably results from early screening of non-affected family members as demonstrated in our present case report.[4] In another study, Steel and associates reported on the clinical features of familial idiopathic interstitial pneumonia, and found that almost 8% of subjects without symptoms of pulmonary fibrosis had HRCT findings consistent with probable or definite interstitial pneumonia.[1] Rosas and colleagues evaluated family members of patients affected by familial IPF to identify asymptomatic subjects with interstitial lung disease (ILD).[3] The cited authors found that 22% of asymptomatic subjects showed radiographic evidence of ILD when screened by HRCT. A consistent finding in previous studies was that a history of cigarette smoking was strongly associated with the development of early ILD in family members of those with familial interstitial pneumonia.[1,3] This suggests that a genetically susceptible individual may develop lung injury from cigarette smoking and exposure to other environmental toxins that may stimulate fibroproliferation, further increasing the risk of developing ILD and progression of pulmonary fibrosis.[1,3,15] In this case report, none of his family member is not suffering from this type of illness. It is possible to speculate that affected individuals might share a single candidate genetic alteration such as a mutation in the surfactant protein C (SP-C) gene (SFTPC), resulting in development of pulmonary fibrosis in our studied family. However, other investigators believe that early onset cases of ILD are distinct, arising from a pathogenic mechanism different from that causing adult onset occurrence of familial IPF.[10] SP-C plays a critical role in reducing surface tension in the alveoli, whereas SP-C absence or mutation in SFTPC mutant patients causes mechanical injury to the respiratory epithelium leading to respiratory failure and severe ILD. This was first reported by Nogee and colleagues who described a mother with desquamative interstitial pneumonia, and her infant with NSIP.[16] Recently, Selman and colleagues evaluated genetic polymorphic variants of the surfactant proteins SP-A1, SP-A2, SP-B, SP-C, and SP-D in a group of patients with IPF.[17] The SP-A16A4 allele was noted to be associated with nonsmoker IPF, whereas the SP-B B1580C allele was more associated with smoker IPF patients compared to control subjects. Thus, in line with previous studies, SPs appear to play an essential role in the pathogenesis of both familial and sporadic IPF. The management of patients with familial IIP is similar to that applied to sporadic cases and is dependent upon the type of IIP diagnosed. However, to date, lung transplantation is the only therapy that has been shown to improve survival in younger individuals with pulmonary fibrosis.

Conclusion

Our report illustrates the importance of obtaining a detailed history and diagnostic role of HRCT of chest in patients with ILD. Though lung biopsy and histopathology is a very useful way to establish the diagnosis and to choose appropriate treatment, this facility is not available in our hospital. Clinical findings, histopathology, and radiographic features are indistinguishable between familial idiopathic interstitial pneumonia and sporadic cases. Younger age at diagnosis is a common finding in patients with the former condition. Cigarette smoking is strongly associated with development of early ILD in family members with familial interstitial pneumonia and both the patients and the family members should be urged to stop smoking.

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