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Evaluation of cases of interstitial lung disease in children

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Abstract

Background: The term childhood interstitial lung disease encompasses a broad group of pulmonary disorders. The present study aimed to evaluate interstitial lung disease in children.

Materials & Methods: The present study was conducted on 78 children age ranged 4-10 years. Patients were divided into two major groups - 'definite ILD' and 'possible ILD' based on their clinical features, results of noninvasive tests such as X-ray and HRCT, and results of invasive tests like bronchoscopy and biopsy. In all patients, clinical feature were recorded.

Results: 42 had definite ILD and 36 had possible ILD. Common clinical features were cough seen in 63 followed by hemoptysis in 41, pallor in 29, clubbing in 28, dyspnea in 26, crepitus and murmur in 7 each. The difference between definite ILD and possible ILD was significant ($P < 0.05$).

Conclusion: Among ILD, definite ILD was seen in 42 and possible ILD in 36. Common clinical features were cough, hemoptysis, pallor, clubbing, dyspnea, crepitus and murmur.

Keywords: clubbing, interstitial lung disease, murmur

Introduction

The term childhood interstitial lung disease (Child) encompasses a broad group of pulmonary disorders that are associated with significant morbidity and sometimes mortality^[1]. Historically, these diseases have been defined based on lung biopsy histopathologic findings. However, recent advances have facilitated increased noninvasive diagnosis through genetic testing and use of chest computed tomography (CT) scans^[2].

The term "interstitial" is, however, misleading, as most of these conditions are associated with abnormalities that are not limited to the lung interstitium but extend to the alveolar and airway compartments. Although it could be argued that "diffuse lung disease" is a better term, the term "child" is in fact now well established in the literature^[3].

The outcome of children with ILD in terms of death and disease-free survival is reported to be 15- 60% and 50%, respectively. The available data on the clinical profile of children with ILD mostly come from small case series that included less than 30 children. Also, many of these reports had focused on one or more specific conditions such as fibrosing alveolitis or desquamative interstitial pneumonitis (DIP) rather than looking at the complete spectrum of ILD^[4].

Children with ILD typically manifest non-specific respiratory signs and symptoms, including tachypnea, hypoxemia, crackles, cough, and poor growth. Because these symptoms overlap those seen in many more common conditions, the first step in diagnostic evaluation is to exclude more common causes of diffuse lung disease (i.e. cystic fibrosis, immunodeficiency, congenital heart disease, pulmonary infection, primary ciliary dyskinesia, and recurrent aspiration)^[5]. The present study aimed to evaluate interstitial lung disease in children.

Materials & Methods

The present study was conducted in the department of Pediatrics. It comprised of 78 children age ranged 4-10 years. The study protocol was approved form institutional ethical committee. Informed written consent was obtained from all parents.

Information regarding name, age, gender etc. was recorded. Patients were divided into two major groups - 'definite ILD' and 'possible ILD' based on their clinical features, results of noninvasive tests such as X-ray and HRCT, and results of invasive tests like bronchoscopy and biopsy. In all patients, clinical feature were recorded. Results were tabulated and subjected to statistical analysis. P value less than 0.05 was considered significant.

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Results

Table I: Distribution of patients

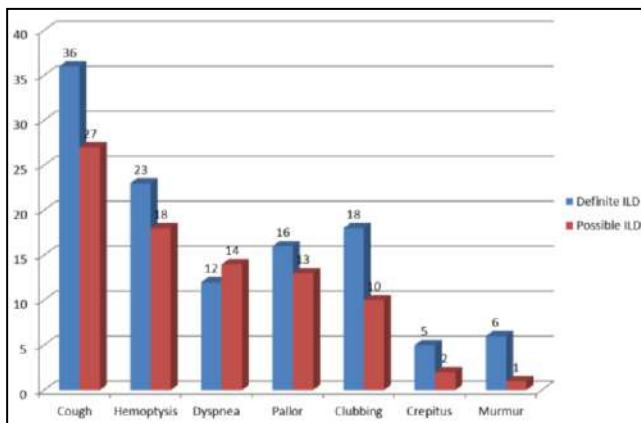
Total- 78		
Types	Definite ILD	Possible ILD
Number	42	36

Table I shows that 42 had definite ILD and 36 had possible ILD.

Table 2: Clinical features in patients

Clinical features	Definite ILD	Possible ILD	Total	P value
Cough	36	27	63	0.01
Hemoptysis	23	18	41	0.02
Dyspnea	12	14	26	0.51
Pallor	16	13	29	0.78
Clubbing	18	10	28	0.01
Crepitus	5	2	7	0.02
Murmur	6	1	7	0.05

Table II, graph I shows that common clinical features were cough seen in 63 followed by hemoptysis in 41, pallor in 29, clubbing in 28, dyspnea in 26, crepitus and murmur in 7 each. The difference between definite ILD and possible ILD was significant (P< 0.05).



Graph I: Clinical features in patients

Discussion

Historically, terminology and classification of interstitial lung disease (ILD) in children have mirrored those of adult disease, but this is generally not helpful. Indeed, there are major differences in disease etiology, natural history, and management between the pediatric age group, in whom ILD most often develops primarily because of an underlying developmental or genetic abnormality, and adults [6].

Chest CT is very useful for defining the extent and pattern of disease with resolution that is superior to plain chest radiographs. Common findings in child may include ground glass opacification, consolidation, and septal thickening. Findings may be suggestive or even specific for some types of ILD, including surfactant dysfunction disorders, bronchiolitis obliterans and Neuroendocrine cell Hyperplasia of Infancy (NEHI) and therefore may reduce need for lung biopsy [7].

Lung biopsy is considered as gold standard for diagnosis of ILD however, getting a lung biopsy in children is difficult especially when they present in advanced stage of illness and have a very high risk for anesthesia. In addition, biopsy may not always be conclusive. Of late, the trend is shifting towards a systematic approach to the diagnosis of patients with ILD rather than subjecting every patient to biopsy. Lung biopsy could possibly be reserved for those children in whom the diagnosis is inconclusive even after noninvasive tests and/or there is poor response to therapy. In children suspected to have ILD secondary to systemic

disorders such as LCH, sarcoidosis etc., a tissue biopsy of the other affected tissues should suffice [9]. The present study aimed to evaluate interstitial lung disease in children.

In present study, out of 78 patients, 42 had definite ILD and 36 had possible ILD. We observed that common clinical features were cough seen in 63 followed by hemoptysis in 41, pallor in 29, clubbing in 28, dyspnea in 26, crepitus and murmur in 7 each.

Paiva MA *et al* [10] conducted a study in which 90 children (median age, 6.8 years; 62% boys) were diagnosed to have ILD during this period. 46 children were classified as having ‘definite ILD’ while 44 had ‘possible ILD’. The commonest clinical features at presentation were cough (82.2%), dyspnea (80%), pallor (50%), and crackles (45.6%). 3 children (3.3%) died while 21 (23%) showed no improvement in clinical status on follow-up at 3 months. A higher ILD score and lower alkaline phosphatase levels were found to be significantly associated with worse outcomes.

The pathogenesis of many forms of ILD remains poorly understood and treatment approaches remain largely empirical. Indeed, there have been no controlled trials of any therapeutic intervention in child. Management is largely supportive, including supplemental oxygen and ventilator support, nutritional support, proper immunizations, and avoidance of harmful environmental exposures. While rheumatologic disorders generally respond well to immunosuppressive medications, there is no clear evidence of efficacy of systemic corticosteroids or hydroxychloroquine in most other forms of childhood ILD [10].

Conclusion

Among ILD, definite ILD was seen in 42 and possible ILD in 36. Common clinical features were cough, hemoptysis, pallor, clubbing, dyspnea, crepitus and murmur.

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