

Fulminant idiopathic pulmonary hemosiderosis associated with coeliac disease: Case report

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Abstract

Lane-Hamilton syndrome refers to the uncommon co-occurrence of idiopathic pulmonary hemosiderosis and celiac disease. We report a case of four years old boy who presented with complaints of respiratory distress and an episode of hemoptysis. He had growth retardation but no diarrheal manifestations of celiac disease. The patient was diagnosed with celiac disease and concomitant idiopathic pulmonary hemosiderosis. He had fulminant progress and needed long term ventilation even though combined immunosuppressive and gluten free diet treatment. But after starting the gluten-free diet, he had no occurrence of episodes of intrapulmonary hemorrhage and severe anemia. As a gluten-free diet has been proven beneficial to the evolution of the coeliac disease, as well as to the respiratory outcome, we recommend a systematic screening for coeliac disease in idiopathic pulmonary hemosiderosis patients.

Keywords

Lane-Hamilton syndrome; gluten free diet; child

Abbreviations

IPH: Idiopathic pulmonary hemosiderosis; CD: celiac disease; LHS: Lane-Hamilton syndrome; GFD: gluten-free diet

Introduction

Idiopathic pulmonary hemosiderosis (IPH) is a rare and serious disease occurs predominantly in infants and children. Clinical symptoms may range from anemia, hemoptysis, dyspnea, hypoxia up to a fulminant course with acute, life-threatening respiratory failure. IPH may occasionally be associated with celiac disease (CD); this association is known as the Lane-Hamilton syndrome (LHS), first reported in 1971 in a young adult [1]. Since then coexistence of IPH and CD has been reported in a limited number of cases [2]. As a gluten-free diet (GFD) has been proven beneficial to the evolution of the celiac disease, as well as to the respiratory outcome of the patients with a LHS.

Case Presentation

A four year old boy presented to us with history of respiratory distress and an episode of hemoptysis. Anamnestic there were no medical problems, despite a pallor and prolonged fatigue. Further, he has a history of pica (eating soil). On physical examination the body weight and height were

13.5 kg and 96 cm (both at 3rd percentile) The heart rate, respiratory rate, blood pressure and oxygen saturation were 142 per min, 68 per min, 90/50 mm Hg and 82% respectively. The patient showed severe pallor. On auscultation, end-inspiratory rales are heard. Investigation showed a hemoglobin level of 2.2 g/dL, leukocytes 14200/cu mm and platelets 382000/cu mm. The mean corpuscular volume was 59 fl, plasma iron 15 µg/dL, iron binding capacity 492 µg/dL and ferritine 20 µg/L. Peripheral smear examination revealed a hypochromic microcytic anemia. Blood gas analyses, serum electrolytes, liver and kidney function tests, coagulation profile, direct Coombs' test and thyroid function test were normal. Chest radiograph showed bilateral diffuse alveolar infiltrates over middle and lower zones, echocardiography was normal. The computer tomography showed some ground glass pattern with bilateral alveolar opacities (Figure). At admission the patient was given empirical antibiotics and blood transfusion. On follow up he got respiratory failure on day 2 and he was ventilated with low tidal volume ventilation (positive end-expiratory pressure of 10 cm H₂O). No infectious or autoimmune diseases could be verified, neither other reasons like Heiners's syndrome, any kind of pulmonary-renal-syndrome or a clotting disorder diagnosis IPH was done. Since an association between CD and IPH has been previously described a workup for CD was also performed. Positive IgA endomysial antibody (1/320 titer) and IgA antigliadin antibody levels were detected. Duodenal biopsy showed partial villous atrophy and increased numbers of intra-epithelial lymphocytes, confirming the diagnosis of CD.

Immunosuppressive therapy with pulses of methylprednisolon (30 mg/kg/day), cyclophosphamide and hydroxychloroquine. There was no marked clinical response to this therapy and transthoracic needle biopsy was performed. The lung biopsy showed haemosiderin laden macrophages in the alveolar spaces. As the serologic findings supported CD, GFD was started. The patient was extubated after 26 days of invasive ventilation and after extubation he was put on non invasive ventilation. He was discharged after a hospital stay of 48 days. Prior to starting GFD he had no occurrence of episodes of intrapulmonary hemorrhage and had gained 2.8 kg weight and 6 cm of height during 1 year of follow-up.

Discussion

Although both CD and IPH are believed to be immunologically mediated, the pathogenetic link between them is not clear. Three pathogenic hypotheses have been suggested in literature to explain the association of IPH and CD: deposition of circulating immune complexes, involving food allergens on the basement membrane of alveolar capillaries; reaction between antireticular antibody and alveolar basement membrane antigen; and an effect of adenovirus 12, a potential causative factor for CD [3].

Idiopathic pulmonary hemosiderosis has significant morbidity and mortality. Progression of pulmonary haemosiderosis is usually lethal and leads to progressive pulmonary fibrosis if left untreated. The clinical interest in this association stems from the fact that treatment of CD with a GFD could lead to remission of IPH. A recent systemic review identified 20 patients with Lane–Hamilton syndrome [4]. Twelve of the 16 patients (75%) prescribed a GFD showed an improvement in the pulmonary symptoms. Our case has fulminant progress and he needed long term ventilation even though combined immunosuppressive and GRD treatment. But after starting the GFD he had no occurrence of episodes of intrapulmonary hemorrhage and severe anemia.

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Several authors have therefore recommended serological screening all patients of IPH for CD. [2,3,5]. The unresolved controversy is whether all patients with IPH should undergo intestinal biopsy or should biopsy be reserved only for those with serologic evidence of CD [6]. The largest series of patients in literature showing response in IPH with GFD has been reported by Khemiri et al. and Sethi et al. [7,8]. Khemiri et al. suggest that screening for CD should be done only in those patients with IPH who have gastrointestinal symptoms [7]. Our patient had growth retardation and chronic anemia, but no diarrheal manifestations of CD. Since half of the cases with LHS in literature had no bowel involvement, we feel that all patients with IPH should undergo both serological testing as well as duodenal/jejunal biopsy.

In conclusion, IPH and CD may have a common pathogenetic link and patients with IPH should be screened for CD even in the absence of gastrointestinal symptoms.

Figure



Figure 1: The computed tomography of the chest demonstrated a patchy, homogeneous opacity with ground-glass pattern in both lungs.

References

1. Lane DJ, Hamilton WS. Idiopathic steatorrhea and idiopathic pulmonary hemosiderosis. *Br Med J* 1971; 2: 89–90.
2. Singhal KK, Janmeja AK, Sodhi R, Punia RS. Hemoptysis in patients of celiac disease with disproportionately severe anemia: tip of the iceberg? *Multidisciplinary Respiratory Medicine* 2013, 8: 25
3. Perelman S, Dupuy C, Bourrillon A. The association of pulmonary hemosiderosis and celiac disease. Apropos of a new case in a child. *Ann Pediatr (Paris)* 1992; 39: 185–188.
4. Agarwal R, Aggarwal AN, Gupta D. Lane-Hamilton syndrome: simultaneous occurrence of coeliac disease and idiopathic pulmonary hemosiderosis. *Intern Med J* 2007; 37: 65-67
5. Paksu S, Paksu MS, Kalayci AG, Sancak R: Lane-Hamilton syndrome: association or coincidence. *Indian Pediatr* 2012, 49(3): 243–244.
6. Pacheco A, Casanova C, Fogue L, Sueiro A. Long-term clinical follow-up of adult idiopathic pulmonary hemosiderosis and celiac disease. *Chest* 1991; 99: 1525–1526.
7. Khemiri M, Ouederni M, Khaldi F, Barsaoui S. Screening for celiac disease in idiopathic pulmonary hemosiderosis. *Gastroenterol Clin Biol* 2008; 32: 745–748.
8. Sethi GR, Singhal KK: Pulmonary diseases and corticosteroids. *Indian J Pediatr* 2008, 75(10): 1045–1056.

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