Idiopathic Pulmonary Hemosiderosis with Celiac Disease; Lane-Hamilton Syndrome

Çölyak Hastalığıyla Birlikte İdiyopatik Pulmoner Hemosiderosiz; Lane-Hamilton Sendromu

IPH with Celiac Disease; Lane Hamilton Syndrome

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Öz

Yirmi bir yaşında erkek hasta acil kliniğimize 2 gündür devam eden kanlı balgam, nefes darlığı, halsizlik ve yorgunluk şikayetiyle başvurdu. Tüm vital bulguları normal olmasına karşın parmak ucu oksijen saturasyonu %92 ölçüldü. Laboratuvar tetkiklerinde hemogramında hemoglobin:8.85g/dL, hematocrit:28.7% olarak tespit edildi. PT ve aPTT normaldi. Romatolojik laboratuvar tetkikleri negatifti. Hafif kanlı bronkoalveolar lavaj sıvısı patoloji preparatlarında "hemosiderin yüklü makrofajlar" görüldü ve idiyopatik pulmoner hemosiderosis (IPH) tanısı konuldu. Gastroduedonoskopi işleminde duedonumda nodülarite görüldü ve bu bölgeden biyopsiler alındı. Biyopsi sonucu çölyak hastalığı ile uyumlu duedonal mukoza villöz atrofisi" olarak rapor edildi. Lane-Hamilton sendromu tanısıyla olguya tedavide sadece glutensiz diyet önerildi. Altı ay takip sonrasında olguda tam remisyon gelişti. Sonuç olarak, çölyak hastalığı ve IPH birlikteliğinden dolayı, IPH'ye bağlı alveoler hemorajilerde, tanı aşamasında uyanık olmamız gerekmektedir. Lane-Hamilton sendromunda glutensiz diyet ile tam remisyon sağlanabilmektedir.

Anahtar Kelimeler

İdiyopatik pulmoner Hemosiderosiz; Çölyak Hastalığı; Lane-Hamilton Sendromu

Abstract

A 21-year-old male patient presented to our emergency department with the complaints of bloody sputum, respiratory difficulty, lethargy, and fatigue persisting for the previous two days. Fingertip oxygen saturation was 92%, while other vital signs were normal. Bilateral ground-glass opacities were present at thoracic computerized tomography. Laboratory findings were hemoglobin:8.85g/dL, hematocrit:28.7%. PT and aPTT values were normal. All rheumatologic laboratory tests were negative. Bronchoalveolar lavage was mildly hemorrhagic and "hemosiderin-laden macrophages" were observed in pathology specimens. The case diagnosed withidiopathic pulmonary hemosiderosis (IPH). Gastroduodenoscopy revealed nodularity in the duodenum, and mucosal biopsies taken from these duedonal regionswere reported as "villous atrophy in mucosal tissues in the duodenum compatible with celiac disease". The only recommended treatment was a gluten-free diet. At follow-up approximately 6 months later, complete remission was achieved. In conclusion, we should be aware, when seeing alveolar hemorrhage related to IPH, that celiac disease can accompany IPH. The concurrence of IPH and celiac disease is known as Lane-Hamilton syndrome. Complete remission in Lane-Hamilton syndrome can be achieved with a gluten-free diet.

Keywords

Idiopathic Pulmonary Hemosiderosis; Celiac Disease; Lane-Hamilton Syndrome

DOI: 10.4328/JCAM.4847 Received: 27.10.2016 Accepted: 28.12.2016 Printed: 01.02.2017 J Clin Anal Med 2017;8(suppl 1): 23-5
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Introduction

Idiopathic pulmonary hemosiderosis (IPH) is a disease of unknown cause that may lead to recurring alveolar hemorrhage. It isgenerally characterized by hemoptysis, iron deficiency anemia, and diffuse parenchymal infiltration in the lungs in childhood. Definitive diagnosis of IPH is made with histopathological observation of hemosiderin-laden macrophages in pulmonary tissue specimens, sputum, or bronchoalveolar lavage BAL [1]. If untreated, IPH may lead to pulmonary fibrosis [2]. Celiac disease (CD) is a malabsorption syndrome, also known as gluten enteropathy, proceeding with chronic diarrhea, weight loss, and findings of delayed growth and development. Inflammation and villous atrophy in the small intestine resulting from autoimmune enteropathy constitute the basic pathophysiology of the disease in genetically predisposed individuals with the digestion of foods containing gluten [3]. Lane-Hamilton syndrome, first described in 1971, is a rare condition representing a combination of IPH and CD [4]. While IPH responds well to corticosteroids, a gluten-free diet can reduce the need for corticosteroids in the treatment of Lane-Hamilton syndrome [5].

Case Report

A 21-year-old male patient presented to our emergency department with the complaints of bloody sputum, respiratory difficulty, lethargy, and fatigue persisting for the previous two days. Physical examination revealed pale sclera and fine rales in the basal areas of both lungs. Fingertip oxygen saturation was 92%, while other vital signs were normal. There was no specific disease or drug in his medical history. Bilateral ground-glass opacities were present at thoracic computerized tomography (CT) (Figure 1).

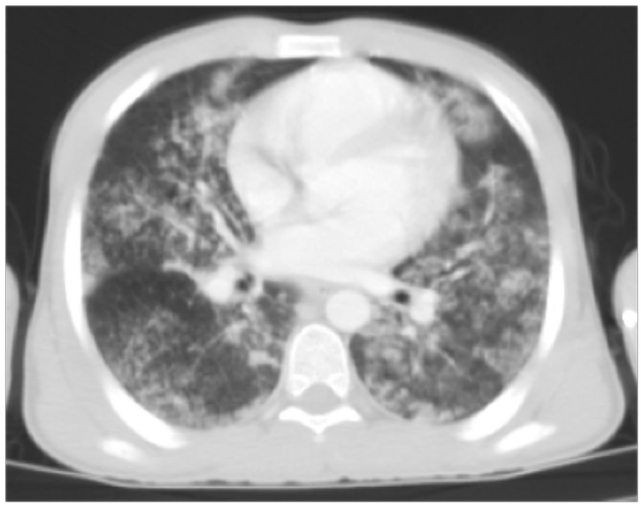

Figure 1. Parenchymal thoracic CT section; hemorrhage-related bilateral ground glass appearance

Laboratory findings were white blood cell:6.24K/uL, red blood cell:3.94 M/uL, hemoglobin: 8.85g/dL, hematocrit: 28.7%, mean corpuscular volume: 72.8 fL, mean corpuscular hemoglobin:22.5pg, mean corpuscular hemoglobin concentration:30.9 g/dL, platelet:355 K/uL, C-reactive protein:0.484 mg/dL, sedimentation:18/hour, glucose:118mg/dL, urea:25 mg/dL, creatinine:0.66 mg/dL, rheumatoid factor<20 IU/mL, iron:12 mg/dL, iron binding capacity:462 mg/dL, total iron binding capacity:464 mg/dL, ferritin: 20.06 ng/ml, vitamin B12:161 pg/mL, and folate:1.9 ng/ml.PT and aPTT were normal. The rheumatologic workup, including all of the antinuclear antibodies, anti-double stranded antibodies, antineutrophil cytoplasmic antibodies, and anti-glomerular basement membrane antibodies were negative.

Fiberoptic bronchoscopy revealed no endobronchial lesions. Bilateral BAL fluid was mildly hemorrhagic. BAL pathology was reported as “hemosiderin-laden macrophages. Therefore, IPH was diagnosed and we treated the patient with corticosteroid (methylprednisolone 1x120 mg) and tranexamic acid (250 mg, 3x1) for five days. The hemoptysis resolved entirely on the third day of treatment and the respiratory symptoms improved dramatically (fingertip oxygen saturation was 96%). The patient, who had hypochromic microcytic anemia, was consulted with the gastroenterology department on the fifth day of treatment. Tests to detect the presence of CD were positive: serum tissue transglutaminase IgA>200 RU/ml, anti-gliadin IgG 200.8 RU/ml, and anti-gliadin IgA >200 RU/ml. Gastroduodenoscopy revealed nodularity in the duodenum, and mucosal biopsies were taken from these duedonal regions (Figure 2A). The biopsies were reported as “villous atrophy in mucosal tissues in the duodenum compatible with celiac disease, focal intraepithelial lymphocyte increase, and crypt hyperplasia with accompanying inflammatory cell infiltration of plasma cells and eosinophils in the lamina propria” (Figures 2B,C,D). CD was diagnosed and we treated the patient with a gluten-free diet. The ground-glass opacities in the pulmonary regions at control thoracic CT resolved in the first week of the treatment. There were no longer any pulmonary symptoms. We discharged the patient on the eighth day of treatment. The only treatment recommendation was a gluten-free diet. At follow-up approximately 6 months later, complete remission was achieved. Hemoglobin levels had returned to normal, and there was no recurrence of bloody sputum or other pulmonary symptoms.

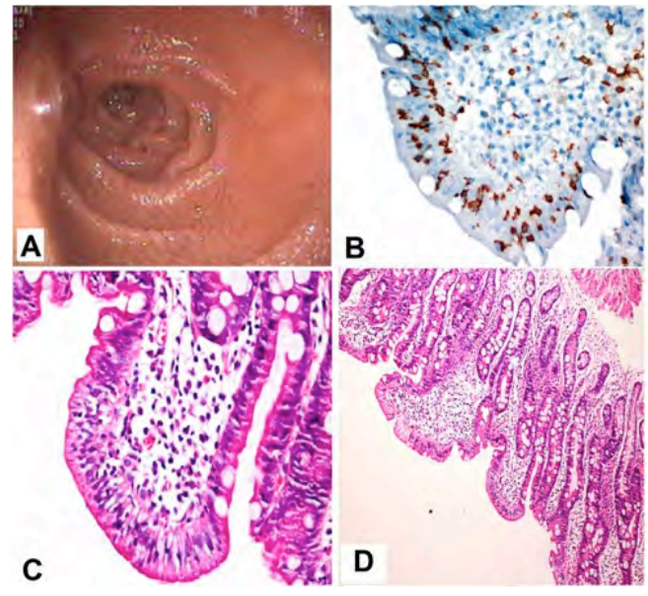

Figure 2. Endoscopic appearance of the duodenum. Appearance of the duodenal mucosa (scalloped feature)(A), Villous athrophy in the small intestine, increased intra-epithelial lymphocytes (hematoxylin-eosin staining) (B), Abundant inflammatory cell infiltration from plasma cells and lymphocytes in the lamina propria (hematoxylin-eosin staining) (C), Increased intra-epithelial lymphocytes staining positive with CD3 (D).

Discussion

Diffuse alveolar hemorrhage (DAH) is a clinical condition involving alveolar hemorrhage deriving from pulmonary microcirculation and alveolar injury. Several conditions involving immune and non-immune mechanisms can lead to DAH. IPH is rarely capable of causing DAH. IPH was first described, as 'brown induration,' by Virchow in 1864 [1]. The prevalence of IPH is estimated at 0.24-1.23/million and IPH is generally seen in adults under the age of 30 [6,7]. A combination of IPH and CD is known as Lane-Hamilton syndrome, the underlying pathogenic mechanism of which is unclear [4]. Parelman et al. implicated three mechanisms in the combination of IPH and CD. The first of these is the accumulation of immune complexes containing food allergens in the basal membranes of alveolar capillaries. The second is cross-relativity between alveolar basal membrane antigen and anti-reticulin antibodies, and the third is the potential involvement of adenovirus 12, a possible etiological agent in celiac disease, in IPH [8]. Studies have reported improvements in pulmonary symptoms associated with IPH following a gluten-free diet used in the treatment of CD [3,5]. In our case, 5-day corticosteroid therapy was administered for the treatment of IPH. After discharge the patient was prescribed a gluten-free diet as treatment. At 6-month follow-up there was no recurrence of either pulmonary or gastrointestinal system (GIS) symptoms with a gluten-free diet, and complete remission was achieved.

In conclusion, we should be aware, when there isalveolar hemorrhage related to IPH, that CD can accompany IPH with or without GIS symptoms. We need to further investigate whether patients with IPHhave GIS symptoms. It should be remembered that in Lane-Hamilton syndrome complete remission can be achieved with a gluten-free diet if the patient is pulmonary asymptomatic, and that corticosteroid therapy can thus be reduced. To the best of our knowledge, there are few case reports about Lane-Hamilton syndrome in the literature. The present case report may point the way to future research about Lane-Hamilton syndrome.

Acknowledgments

The authors wish to thank Recep Tayyip Erdoğan University Medical Faculty staff in Rize for their assistance.

Competing interests

The authors declare that they have no competing interests.

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How to cite this article:
Karataş M, Özyurt S, Gümüş A, Kara BY, Şahin Ü. Idiopathic Pulmonary Hemosiderosis with Celiac Disease; Lane-Hamilton Syndrome. J Clin Anal Med 2017;8(suppl 1): 23-5.