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PULMONARY HEMORRHAGE TREATED WITH GLUTEN FREE DIET: LANE HAMILTON SYNDROME

Internal Medicine

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ABSTRACT

Lane Hamilton syndrome can be a challenging diagnosis when the patient has no gastrointestinal symptoms. We present a case of a 20-year-old male who presented with shortness of breath, hemoptysis and severe iron deficiency anemia but did not have any symptoms of celiac disease. We would like to highlight that serological markers for celiac disease should be sought in all patients with Idiopathic Pulmonary Hemosiderosis. All the symptoms in these patients can be successfully treated with gluten free diet.

KEYWORDS

Celiac disease, Idiopathic Pulmonary Hemosiderosis, serological markers, gluten free diet

INTRODUCTION

Idiopathic pulmonary hemosiderosis (IPH) is a rare disorder with an incidence of 0.24 and 1.23 per million children per year. [1] The association of IPH with celiac disease (CD) is even rarer and it is called Lane-Hamilton Syndrome. We report a case of Lane Hamilton Syndrome in an adult who presented with the classical triad of IPH that is hemoptysis, severe iron deficiency anemia and radiological evidence of pulmonary infiltrates but did not have any symptoms of celiac disease. We would like to highlight the rarity of this condition especially in adults and the importance of checking serological markers of celiac disease even in the absence of celiac symptoms as a simple intervention like gluten free diet is shown to improve the clinical status of the patient

Case Presentation

A 20-year-old man with no past medical history presented to the emergency department with shortness of breath, palpitations and lightheadedness for 3 weeks and occasional hemoptysis for one year. On examination, the patient was tachycardic with a HR of 104 beats/min and BP of 129/71 mm Hg. Laboratory findings were significant for a hemoglobin (Hb) of 4.1 g/dL with a hematocrit (Hct) of 14.5%, reticulocytes were elevated at 5.8%, haptoglobin of 165 mg/dL, ferritin of 42 ng/mL, lactate dehydrogenase of 209 U/L, iron level of 13 ug/dL, total iron binding capacity of 414 ug/dL. Peripheral smear was normal and fecal occult stool blood test was negative. He was administered 4 units of PRBCs.

As hemolysis labs were negative, further work up of iron deficiency anemia was sought. Computed tomography of the thorax with contrast was sought for hemoptysis which showed diffuse ground glass opacities in bilateral lungs with relatively sparing of the apices suggestive of either pulmonary hemorrhage or atypical pneumonia as shown in figure 1. Bronchoscopy showed diffuse alveolar hemorrhage and transbronchial biopsies showed intra-alveolar hemosiderin. Infectious work up of the bronchoalveolar lavage including fungal culture, general viral culture, Legionella, Pneumocystis Jerovici, acid fast staining for Tuberculosis, HIV were negative. Work up for vasculitis was negative, however celiac panel was positive with tissue trans-glutamin IgA of 100 and Gliadin of 84. The patient was diagnosed with Lane Hamilton syndrome and was counseled extensively on adapting a gluten free diet. The patient was followed up 6 months and one year later and his pulmonary symptoms completely resolved on gluten free diet.

DISCUSSION

The first case of Lane Hamilton syndrome was described in 1971 by Lane D J et al. [2] The pathogenetic link for the unusual association of celiac disease and IPH is unknown and there is an ongoing debate on if the occurrence of these two conditions is just a co-incidence. Celiac disease is an enteropathy characterized by life-long intolerance to ingested gluten in genetically susceptible individuals whereas IPH presents as hemoptysis and diffuse pulmonary infiltrates. [3] Several hypotheses are proposed for the association between the two conditions. CD 8+ T lymphocytes play a major role in the pathogenesis of celiac disease. It is observed that an increase in the ratio of CD4/CD8 was seen in the bronchoalveolar fluid analysis. Two other hypotheses were a cross-reaction between alveolar basal membrane antigen and anti-reticulin antibodies and adenovirus 12, the potential etiological factor of CD is thought to also play a role in IPH. [4]

Most patients present with only one of the features of the triad of IPH. The gold standard for diagnosing IPH is with a lung biopsy but it can also be diagnosed when hemosiderin-laden macrophages are found in the bronchoalveolar lavage. Given the rarity of the condition and the variation and severities in presentation, a mean delay of 2.4 years was noted in the diagnosis of IPH in Romanian experience and 30 months in Indian experience. [7] As Lane Hamilton syndrome is even rarer, the diagnosis is further delayed especially when patients do not have the gastrointestinal symptoms of gluten intolerance. Hence, it is now strongly recommended to perform serological testing for CD in patients with IPH even in the absence of GI symptoms. It is an unresolved controversy if patients with IPH should undergo an intestinal biopsy for the confirmation of celiac disease. [4] [5] [6] [8]

It is of profound interest that the treatment of celiac disease with gluten free diet also resolves pulmonary symptoms. [3] In a case series reported by Gulshan et al., all 3 children with Lane Hamilton showed improvement within 8-17 months of dietary changes. Steroids and immunosuppressants could be weaned off in these patients. [5] Lane-Hamilton syndrome is a well-documented syndrome but there is paucity of literature on the pathogenesis, diagnosis and treatment of this condition.

CONCLUSION

Diagnosis of Lane-Hamilton syndrome can be missed easily leading to long-term of use of steroids and immunosuppressants when these patients can be successfully treated with dietary changes, an intervention with no side effects. We recommend testing for serological markers of celiac disease in all patients with IPH irrespective of presence of symptoms of gluten intolerance.

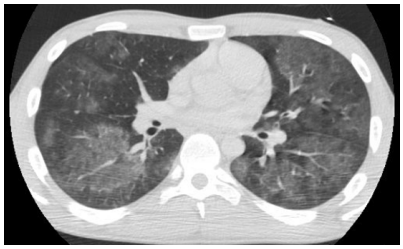

Figure 1: Computed tomography imaging of the thorax showing diffuse ground glass opacities in bilateral lungs

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