Late presentation of lane-hamilton syndrome in a 33 year old female: A case report

Abstract

Lane-Hamilton syndrome (LHS) is a rare syndromic association between idiopathic pulmonary hemosiderosis and celiac disease (CD). It is usually seen in children below 15 years of age. It can occasionally be seen in adults. We present the case of a 33-year-old female patient who presented with recurrent episodes of hemoptysis to the pulmonary outpatient department. She also gave a history of having frequent loose stools. She was admitted and investigated thoroughly and was found to be suffering from LHS which is a rare disease. High-resolution computed tomography (HRCT) of the chest and duodenal biopsy helped in concluding the diagnosis. She was started on gluten-free diet (GFD) and has responded well with no episodes of hemoptysis on 9-month follow-up and is in good general condition. This case highlights the importance of keeping a high index of suspicion of LHS in a young patient presenting with unexplained hemoptysis and diarrhea. In a known case of CD presenting with hemoptysis, a HRCT chest aids in the diagnosis of LHS. A GFD is the mainstay of long-term treatment, and adherence to this diet shows remarkable improvement in the symptoms of the patient and their overall general condition.

Keywords: *Case report, diarrhea, hemoptysis, Lane-Hamilton syndrome*

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Introduction

Lane-Hamilton syndrome (LHS) is defined as the association of celiac disease (CD) and idiopathic pulmonary hemosiderosis (IPH). They are both immunologically mediated diseases with no clear pathogenic link between them. They are usually seen in children < 15 years of age and occasionally in adults. The patients usually presented in poor general condition, stunted growth, malnutrition, clubbing of nails, frequent diarrheal episodes, cough, and hemoptysis. Patients usually benefit from the gluten-free diet (GFD) and show marked clinical improvement.

Case Report

We present the rare case of a 33-year-old thin and lean married housewife female who presented to the pulmonary outpatient department (OPD) with recurrent episodes of hemoptysis, passing frequent loose stools, and chronic malabsorption with recent worsening of the symptoms, which prompted her to seek medical care. There was no significant past medical or family history. There is no significant psychosocial history. She did not seek any medical treatment before the present episode.

On examination, she was severely anemic, malnourished, and had a low body mass index. She had hyperdynamic circulation. She had clubbing of nails, pale conjunctiva, and was irritable.

She was investigated and the workup was done for autoimmune and infective etiologies, which was negative. A high-resolution computed tomography (HRCT) of the chest was done to evaluate for episodes of hemoptysis, which showed ground-glass attenuation in bilateral lung parenchyma with diffusely scattered small ground-glass nodules [Figure 1]. Bronchoalveolar lavage (BAL) and transbronchial lung biopsy were done. BAL fluid cytocentrifuge smears were adequate and showed numerous pigment-laden alveolar macrophages. More than 90% of alveolar macrophages showed Prussian blue cytoplasmic granules on Perl's stain, confirming hemosiderin [Figure 2a]. Transbronchial lung biopsy showed largely maintained pulmonary architecture and intra-alveolar aggregates of

How to cite this article: Mishra SC, Gupta A, Sharma S, Gupta A. Late presentation of lane-hamilton syndrome in a 33 year old female: A case report. Int J App Basic Med Res 2022;12:137-9.

Submitted: 21-Sep-2021
Revised: 15-Feb-2022
Accepted: 14-Mar-2022
Published: 10-May-2022

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Access this article online

Website: www.ijabmr.org

DOI: 10.4103/ijabmr.ijabmr_620_21

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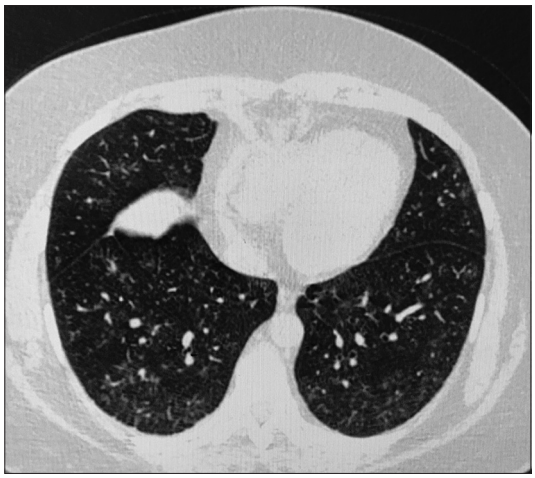

Figure 1: Axial chest high-resolution image showing ground-glass attenuation in both lungs

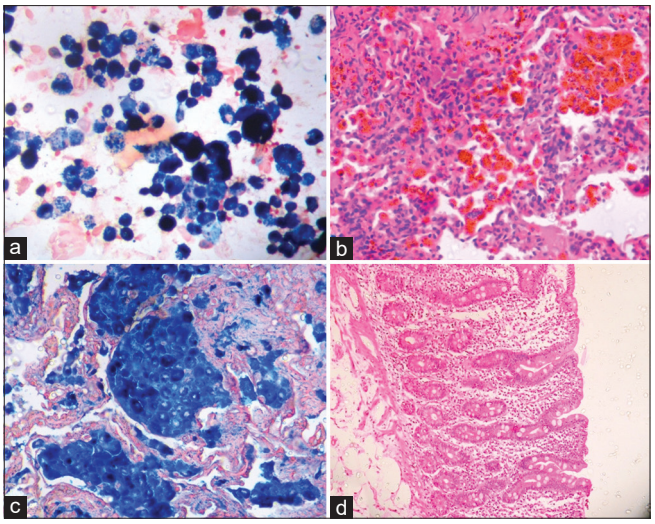

Figure 2: (a) ×400 magnification: Bronchoalveolar lavage cytocentrifuge smears on Perl's stain show numerous hemosiderin-laden macrophages (hemosiderin appears Prussian blue on Perl's stain). (b) ×400 magnification: transbronchial lung biopsy shows unremarkable pulmonary architecture, and alveoli are filled with aggregates of pigment-laden macrophages. (c) ×400 magnification: Perl's stain in transbronchial lung biopsy shows Prussian blue aggregates of hemosiderin-laden macrophages. (d) ×100 magnification: duodenal biopsy shows partial villous atrophy with crypt hyperplasia

hemosiderin-laden macrophages (HLM) [Figure 2b]. These macrophages showed Prussian blue cytoplasmic granules on Perl's stain, consistent with hemosiderosis [Figure 2c]. Given the history of recurrent episodes of loose stools, an anti-tissue transglutaminase test was done, which came out to be positive. She then underwent endoscopy and biopsy of the second part of the duodenum. Endoscopic duodenal biopsy showed partial villous atrophy, crypt hyperplasia, and increased intraepithelial lymphocytes, consistent with celiac spruce [Figure 2d]. Corroborating the findings of HRCT chest, BAL fluid centrifuge microscopy, and transbronchial lung biopsy tissue histopathology with the clinical presentation, a diagnosis of LHS was concluded.

She was then treated for CD with a GFD, to which she responded very well and has not had an episode of hemoptysis for the last 9 months. She also started on oral steroids. Anemia was treated with iron supplements and an iron-rich diet.

She is on follow-up in the pulmonary department OPD and is doing well clinically.

Discussion

Association of IPH and CD is known as LHS. It is believed that both these disorders are immunologically mediated,[1,2] However, the causal relationship is still not clear,[2-5] IPH mainly affects adolescents and children and is an infrequent disease with an incidence of around 0.24–1.23 patients/million children,[6,7] A triad of recurrent alveolar hemorrhage, hemoptysis, and iron-deficiency anemia raises suspicion of IPH and BAL confirms the diagnosis by showing HLM in BAL fluid.[8] However, lung biopsy is considered the gold standard for diagnosis. Because of the syndromic association of CD and IPH, many authors recommend screening of all the patients of IPH for CD even in the absence of gastrointestinal symptoms.[3,4] This is similar to our case where the patient presented primarily for distressing pulmonary symptoms and she was investigated for the CD with serological testing, endoscopy, and duodenal biopsy and the syndromic affliction could be ascertained.

A GFD for CD, along with or without steroids or immunosuppressants, and iron therapy, is the mainstay for the treatment of LHS. The use of GFD is known to result in ceasing lung hemorrhage, decreased requirement for blood transfusions, and improvement in lung function in cases of LHS.[3-5]

Our patient has responded very well to a combined approach using steroids, oral iron supplements, and a GFD and has had no episodes of hemoptysis at 9 months follow-up.

Conclusions

All the young patients with recurrent episodes of hemoptysis and chronic malabsorption should be evaluated for LHS. Especially in the clinical setting where the anemia severity is disproportionate to the radiologic findings. As both components of the LHS, i.e., IPH and CD benefit greatly from a GFD, all the patients of IPH should be assessed for CD.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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