Correspondence

To the Editors

Re: case report "Idiopathic pulmonary haemosiderosis: A rare cause of anaemia"

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(Key words: Idiopathic pulmonary haemosiderosis; anaemia)

I read with interest the case report titled "Idiopathic pulmonary haemosiderosis: A rare cause of anaemia"[1]. The authors have described two children with anaemia due to pulmonary haemorrhage of obscure cause. Although the workup described in both patients has been extensive, a few crucial things in the workup of a child with idiopathic pulmonary hemosiderosis (IPH) need to be considered.

I wish to clarify the terms 'pulmonary haemorrhage' and 'pulmonary haemosiderosis', which are often used interchangeably. 'Pulmonary haemorrhage' denotes bleeding into the lungs and the conducting airways. 'Pulmonary haemosiderosis' denotes the presence of lung macrophages containing the degradation products of haemoglobin, haemosiderin-laden macrophages. Thus, pulmonary haemosiderosis is a pathologic state caused by bleeding of any type into the lungs[2]. The diagnosis of IPH hinges on establishing diffuse pulmonary haemorrhage and an extensive workup to rule out secondary causes of haemosiderosis. IPH is a diagnosis of exclusion.

In both patients prothrombin time and/or activated partial thromboplastin time have not been reported. Therefore a coagulopathy cannot be ruled out as a cause of pulmonary bleed. Workup for complement levels (C3, C4, CH50) could have indicated an immune complex disease.

In the first patient antinuclear antibody, anti-glomerular basement membrane antibody and anti-neutrophil cytoplasmic antibody should have been done. Even a urine microscopy could have indicated the presence of associated nephritis in this patient.

Second patient presented with pallor, decreased appetite and weight loss for 7 months and febrile illness for 2 weeks. Bronchoalveolar lavage (BAL) in this patient did not reveal any evidence of infection. Significant weight loss, decrease in appetite and febrile illness make intensive workup for an infectious cause imperative before diagnosing IPH. Was a workup for human immunodeficiency virus and primary immunodeficiency done in this patient? Did the patient receive antibiotics with steroids or only steroids? In India epidemiologically tuberculosis (TB) would be a more relevant cause for secondary haemosiderosis in this patient. Being a pauci-bacillary disease in children, microbiological diagnosis is challenging. BAL, despite being negative for acid fast bacilli on smear, can show growth of mycobacterium tuberculosis on culture media. Was an effort to find corroborative evidence of TB by radiological chest survey of contacts or purified protein derivative test in the patient made?

An unequivocal diagnosis of pulmonary haemosiderosis can often be made by a combination of clinical and serological findings and BAL fluid examination[3]. Thus a lung biopsy should not be attempted without complete serological investigations, renal functions tests and urinalysis.

Most significantly, the authors have not indicated workup for coeliac disease in both patients. There have been a number of reports indicating association of coeliac disease with IPH, also known by the eponym Lane-Hamilton syndrome. Presence of associated coeliac disease has a major therapeutic implication in patients with IPH. Coeliac disease is a multisystem, immunologically mediated disease elicited by the ingestion of gluten in genetically susceptible persons. The treatment is lifelong dietary omission of gluten, which is present in wheat, barley, and rye. There have been a number of reports indicating remission in pulmonary symptoms with gluten free diet in Lane Hamilton syndrome[4].

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Response by authors of article

We very much appreciate Dr. Singhal's feedback on our case report. Both patients had been thoroughly investigated but all the investigations were not mentioned due to limitations on the word count. Prothrombin time, activated partial thromboplastin time and urine microscopy were normal in both patients. We intended to do antinuclear antibody, anti-glomerular basement membrane antibody and anti-neutrophil cytoplasmic antibody even in the first patient but due to financial issues it could not be done. Moreover, lung biopsy (light microscopy, electron microscopy and immunofluorescence studies) was not evident of autoimmune disease.

Bronchoalveolar lavage in case of both patients was sent for microscopy as well as bacterial, fungal, and mycobacterial culture studies and the culture reports were negative. Tuberculin test was also negative. HIV by ELISA was nonreactive in both our patients. Work-up of primary immunodeficiency is to be done if patient fulfils criteria for its work-up (given in Nelson's textbook of Pediatrics). Moreover the patients did not have a severe infection or other features suggesting a primary immunodeficiency. Regarding, Lane-Hamilton syndrome both IPH and coeliac disease conditions are considered to be immune-mediated, although the causal relationship is not clear[1,2,3]. Studies showed that the two conditions may be associated in 6.6-8.7% cases[4,5]. Hence the anti TTG antibodies were sent in the second patient also keeping in mind loss of appetite and weight loss and were found to be negative.

We attempted the lung biopsy only after thorough serological investigations as lung biopsy is the gold standard to confirm the diagnosis of pulmonary haemosiderosis[6].

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