International Journal of Contemporary Pediatrics
Krishnasamy J. Int J Contemp Pediatr. 2017 Jan;4(1):272-273
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291
DOI: http://dx.doi.org/10.18203/2349-3291.ijcp20164617

Case Report

Childhood coeliac disease presenting as hepatitis with an associated aplastic anaemia: a case report and literature review

Jagannathan Krishnasamy*

Department of Pediatrics, GKNM Hospital, Coimbatore, Tamilnadu, India

Received: 16 September 2016
Accepted: 21 October 2016

***Correspondence:**
Dr. Jagannathan Krishnasamy,
E-mail: jagannathkrish@gmail.com

ABSTRACT

Although aplastic anaemia and deranged liver function test are well documented in the adult coeliac literature, this combination is not well recognized in childhood. We report a case of 8 year old girl who presented with jaundice and spontaneous bruising. Initial investigations showed pancytopenia, raised bilirubin and transaminases. On extensive evaluation to look in to the cause of above presentation, she was found to have coeliac disease with aplastic anaemia. She was started on gluten free diet which resolved her liver derangement. The marrow aplasia persisted however, requiring immune therapies.

Keywords: Aplastic anaemia, Coeliac disease, Deranged liver function test, Pancytopaenia

INTRODUCTION

Coeliac disease (CD) is gluten induced immune disorder in genetically susceptible individuals with characteristic clinical manifestations, CD-specific antibodies, HLA-DQ2 or HLA-DQ8 haplotypes, and enteropathy.[1]

CD clinical manifestations are divided into two types: the classic type presents with chronic diarrhoea, abdominal distension, weight loss and coeliac crisis. The non-classic type divided into gastrointestinal manifestations which include recurrent abdominal pain, growth delay, vomiting, poor appetite and constipation, and extra-gastrointestinal manifestations include pubertal delay, dental enamel defects, irritability, anaemia, headache, vertigo, aphthous stomatitis, dermatitis herpetiformis, osteoporosis, arthritis, neurologic symptoms and unexplained elevation of transaminase levels.[2,3] Therefore, diagnosing coeliac disease requires a high degree of suspicion, followed by correct screening and a confirmatory test with an intestinal biopsy. Study was presenting a case of childhood coeliac disease with aplastic anaemia and liver involvement.

CASE REPORT

An eight year old girl presented with jaundice and spontaneous bruising. The episode was preceded by gastroenteritis for a week. She did not have any long term gastrointestinal symptoms. Examination revealed jaundice, scratch marks, petechiae and mild hepatomegaly. The initial blood test showed haemoglobin of 11.6 g/dL, white cell count of 2.6 x 109/L and platelets of 6 x 109/L. The total bilirubin was 7.2mg% with the direct fraction of 6.1 mg%, AST 1028 u/L, ALT 1358 u/L, alkaline phosphatase 300 u/L and gamma glutamyl trans peptidase of 95 u/L. INR was 1.5 with albumin being 3.9 g/dL. The bone marrow aspiration showed hypocellular marrow. She had extensive liver work up. Ammonia, lactate, caeruloplasmin, alpha-fetoprotein and alpha-1-antitrypsin phenotype were all within normal limits. Immunoglobulins, complement and autoimmune screening were normal. The hepatitis serology including hepatitis A, B, C and E, HIV, cytomegalvirus, Ebstein-Barr Virus and adenovirus were all negative. Surprisingly anti-tissue transglutaminase was raised and it was more than 128 U/ml. Because of the rarity of the presentation,

she did have further testing. The anti-endomysial antibody was positive and HLA screening showed DQ2 positivity. Following endoscopy, the histopathology from the duodenum showed intraepithelial lymphocytosis and loss of villi, which confirmed coeliac disease.

She was advised gluten free diet, vitamin K and ursodeoxycholic acid. Her bilirubin and transmaminitis gradually improved and then normalised after commencement of gluten free diet. Ursodeoxycholic acid and vitamin K was stopped. For aplastic anaemia, she received packed cell transfusions and platelet transfusions. She was treated for neutropaenic sepsis with intravenous antibiotics. She needed immune therapy for her aplastic anaemia. She received one course anti-thymocyte globulin and steroids. She was on cyclosporine A. Her neutrophil count and platelet count improved and she had good reticulocyte response. T-cell subset, especially CD4 count was done regularly to guide in weaning the cyclosporine A. She is under regular follow up with the haematology and paediatric team.

DISCUSSION

There are not many paediatric cases with CD and aplastic anaemia, even though there are adult case reports. Maheswari et al, reported a 13 year boy who presented with bilateral knee and ankle pain with pancytopaenia and haematochezia. He presented with short stature. The diagnosis of coeliac disease was established due to strongly positive anti-tissue transglutaminase antibody in the serology and villous atrophy with intraepithelial lymphocytosis in the duodenal biopsies. Bone marrow examination revealed hypocellular marrow. He was commenced with gluten free diet. He received antibiotics, packed cell transfusions and platelet transfusions. The blood cell counts improved over a period of 8 months and the response sustained. He did not need any immunosuppression.[4] Badyal et al reported a child with coeliac disease who was not under strict gluten free diet, developed aplastic anaemia.[5]

Grey-Davies adult case series reported three patients who presented with aplastic anaemia associated with coeliac disease. One of them had deranged liver function. All of them were found to have positive coeliac serology and positive HLA-DQ2. Eventually the diagnosis was confirmed by small bowel biopsy. Bone marrow examination revealed hypocellular marrow. All of them were commenced on gluten free diet and they were all treated or planned for anti-thymocyte globulin and cyclosporine A.[6]

In another adult case series, 5 patients were included in the study. Three patients were diagnosed to have coeliac disease at the same time as aplastic anaemia. In two patients, coeliac disease was diagnosed before aplastic anaemia. Four of these patients were HLA-DQ2 positive. All patients were treated with anti-thymocyte globulin and cyclosporin A. Only one patient responded to this treatment. 3 patients underwent haematopoietic stem cell transplant.[7]

Study patient is the first paediatric case to present with combination of coeliac disease, aplastic anaemia and deranged liver function. She presented with pancytopaenia, raised bilirubin and transaminases. She was planned for bone marrow transplant from a blood related donor, but it was withheld considering the improving clinical condition as the marrow started to work.

CONCLUSION

Coeliac disease should be considered in newly diagnosed aplastic anaemia patients. Even though there are not many case reports, extrapolating the evidence from the adult case series, the paediatric population may be subjected to anti-thymocyte globulin and cyclosporine A treatment for aplastic anaemia before considering bone marrow transplant.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Husby S, Koletzko S, Korponay-Szabo IR, Mearin ML, Phillips A, Shamir R, et al. European Society for Pediatric Gastroenterology, Hepatology, and Nutrition Guidelines for the Diagnosis of Coeliac Disease. JPGN. 2012;54(1):136-60.
2. Cozzali R, Castellucci G, Carosati E, Dominijanni V, Ferraro L. Changes in pediatric presentation of celiac disease. Digestive and Liver Disease. 2013;45:e281-282.
3. Guandalini S, Assiri A. Celiac disease: a review. JAMA Pediatr. 2014;168(3):272-8.
4. Maheshwari A, Nirupam N, Aneja S, Meena R, Chandra J, Kumar P. Association of celiac disease with aplastic anemia. Indian J Pediatr. 2012;79(10):1372-3.
5. Badyal RK, Sachdeva MUS, Varma N, Thapa BR. A rare association of celiac disease and aplastic anemia: case report of a child and review of literature. Pediatr Develop Pathol. 2014;17(6):470-3.
6. Grey-Davies E, Hows JM, Marsh JC. Aplastic anaemia in association with coeliac disease: a series of three cases. British J Haematol. 2008;143(2):258-60.
7. Salmeron G, Patey N, de Latour RP. Celiac disease and aplastic anemia: a specific entity. British J Hematol. 2009;146:113-24.

Cite this article as: Krishnasamy J. Childhood coeliac disease presenting as hepatitis with an associated aplastic anaemia: a case report and literature review. Int J Contemp Pediatr 2017;4:272-3.