Journal of Advances in Medicine and Medical Research

23(5): 1-4, 2017; Article no.JAMMR.35609
ISSN: 2456-8899
(Past name: British Journal of Medicine and Medical Research, Past ISSN: 2231-0614, NLM ID: 101570965)

An Unusual Presentation of a Primaryeosinophilic Gastrointestinal Disorder (Case Report)

M. C. Fadous Khalife[1*], P. Noun[1], F. Semaan[1], R. Dagher[1], M. Karam[2], W. Ghaname[1], R. Ghiye[1], R. Chaabane[1], M. Samarani[1] and M. Al-Ojaimi[3]

[1]Faculty of Medicine, Holy Spirit University of Kaslik, UH Center of Notre Dame Des Secours, Kaslik, Lebanon.
[2]American University Hospital of Beirut, Beirut, Lebanon.
[3]Balamand University, Koura, Lebanon.

Authors' contributions

This work was carried out in collaboration between all authors. Authors MCFK, PN, FS and MAO designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors MCFK, MK, WG and MAO managed the analyses of the study. Authors MCFK, RG, RC and MS managed the literature searches. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/JAMMR/2017/35609
Editor(s):
(1) Ige, Serah Funke, Department of Physiology, Faculty of Basic Medical Sciences, College of Health Sciences, Ogbomoso, Nigeria.
Reviewers:
(1) Tolga Dinc, Ankara Numune Training Hospital, Turkey.
(2) Jagtap Sunil, Krishna Institute of Medical Sciences University, India
Complete Peer review History: http://www.sciencedomain.org/review-history/20569

Case Report

Received 21st July 2017
Accepted 11th August 2017
Published 19th August 2017

ABSTRACT

Primary Eosinophilic gastroenteritis is an uncommon inflammatory gastrointestinal disease affecting children first described in 1937.Its incidence is 1/100000. The eosinophilic infiltration can occur in one or more segments of the gastrointestinal tract especially stomach and duodenum in the absence of known etiology. One or more layers are infiltrated by eosinophils. The clinical manifestations are heterogeneous. Its pathogenesis is not fully understood but food hypersensitivity is considered a major factor. Treatments are mostly unsatisfactory, and prognosis is uncertain. No guidelines up to this date have been published regarding the management of these patients.

We report the case of a premature twin boy who was admitted to the –Neonatal Intensive Care Unit-NICU for suspected sepsis.

**Corresponding author: E-mail: mclaude454@hotmail.com;*

Our purpose is to highlight the importance of multidisciplinary team approach for management of unusual neonatal cases where sub-specialized workup and skills are a must for early diagnosis and clinical success. Our case is peculiar because of the young age of presentation and the scarce gastrointestinal symptoms at presentation that swayed us towards a hematologic or infectious disorder which are the likely diagnoses at this age.

Keywords: Eosinophilic gastro-enteritis; hypersensitivity; inflammatory disease; sepsis.

1. INTRODUCTION

Primary Eosinophilic gastroenteritis is an uncommon inflammatory gastrointestinal disease affecting children first described in 1937 [1]. Its incidence is 1/100000 [2]. The eosinophilic infiltration can occur in one or more segments of the gastrointestinal tract – GI-especially stomach and duodenum in the absence of known etiology. In some cases, the esophagus and colon are involved. One or more layers are infiltrated by eosinophils. The clinical manifestations are heterogeneous based on the location of the eosinophilic infiltrates and the depth of the GI involved. The usual clinical presentation includes: abdominal pain, nausea, vomiting, diarrhea and weight loss.

The pathogenesis of this disorder not fully understood but food hypersensitivity is considered a major factor. Patients often have a history of atopy or food allergies.

Treatments are mostly unsatisfactory, and prognosis is uncertain. No guidelines up to this date have been published regarding the management of these patients.

We hereby report the case of a 3 days old who presented with decreased oral intake, leukocytosis and eosinophilia. He was diagnosed with Primary Eosinophilic gastroenteritis at 10 months of age.

Our main purpose is to raise awareness about this rare entity and highlight the importance of multidisciplinary team approach for the management of unusual neonatal cases where sub-specialized workup and skills are a must for early diagnosis and clinical success.

2. CASE REPORT

A 36 weeker baby boy member of a twin gestation - twin B- with an uneventful pregnancy course and a smooth delivery; was admitted to the NICU at day three of life for decreased oral intake. Twin brother A was admitted at 6 hours of life for pneumonia. Birth weight 2010 g.

His blood tests showed an elevated white blood count (WBC) of 17700/microliter (neutrophils 47.9%, lymphocytes 28%, monocytes 13% and eosinophils 9%: absolute number 1.66 K/µL), C-reactive protein (CRP) of 82 mg/L and procalcitonin 3.93 ng/ml. An empiric antibiotic therapy of ampicillin/cefotaxime for 10 days was given although the blood, urine and cerebral spinal fluid (CSF) cultures were negative. We noticed an amelioration of his feeding pattern after 48 hours, his CRP dropped to 17mg/l and procalcitonin to 0.355 ng/ml. He was discharged home against medical advice after completing the 10 days of antibiotics with WBC 27700/uL (N 25%, L 35,3%, Eos 31,7%, absolute number 8.78 K/µL) CRP 2 mg/dl.

Two days after discharge, a follow up CBC revealed: WBC count 39000 /ul (N 25%, Eos 35. 8%absolute count 9.9K/µL) and CRP 11mg/l. He was admitted to another facility and started on antibiotics. The patient was clinically deteriorating with worsening respiratory distress requiring intubation. Chest X-Ray done was normal. Patient was given 25 days of high potency antibiotics and multiple regimens were tried to no avail- CRP 49 mg/L. He was extubated on day 6. A total body scan was done and revealed no abnormalities. He received a transfusion of packed Red blood cells since Hb=6 g/dl. All antibiotics were stopped and patient was discharged from NICU at age 40 days on Neocate pending further diagnostic workup.

At the age of 2 months 18 days, he was hospitalized again in our service because of fever 39℃ for 24 hours' duration. His weight was 4.5 kg. Complete Blood Count –CBC- done revealed WBC 21100/ µl (26% neutrophils, 43% lymphocytes, 15% monocytes and 14% eosinophils AC 3.8K/µL), Hb 8g/dl and MCV 83fl. CRP was 24 mg/dl, ESR 95 mm at the first hour, ferritin 544.66 ng/mL with a procalcitonin of 0.157 ng/mL. Viral serology for cytomegalovirus, Epstein-Barr virus, parvovirus B19 and herpes virus were negative. Stool parasitology test was also negative. Flow cytometry of peripheral blood and bone marrow cells showed no abnormalities.

The level of immunoglobulin was normal for age: IgA, IgG and IgM with IgE level =10.09 IU/mL. Hemoglobin electrophoresis was normal. Patient was discharged home on Esomeprazole 10 mg /day because of recurrent vomiting. Patient had seborrheic dermatitis and facial atopic dermatitis. He was having watery stools in spite of Neocate.

At age 7 months he was admitted for investigations of poor oral intake, his chronic diarrhea and inadequate weight gain. Any attempt to introduce solid food failed due to worsening of his vomiting. Mother reported one episode of bilateral acute otitis media-AOM- and another of left ruptured AOM in the interim period. Due to persistent leukocytosis (WBC 26500/µL, 32% neutrophils, 47% lymphocytes, 6% monocytes, 12% eosinophils AC 3.2), anemia and elevated ESR (83 mm for the first hour); more advanced investigations were sought, like Janus Kinase 2 (JAK-2) mutation, Complements C3 and C4, Anti-nuclear antibody (ANA), anti- double stranded DNA (anti-DNA), anti- neutrophil cytoplasmic antibodies (C-ANCA and P-ANCA) which turned out negative. Ultrasound of abdomen revealed no abnormalities. Total body MRI showed minimalthoraco-abdominal adenopathies. Other laboratory tests were repeated, the dosage of folate, vitamin B12, iron level and total iron binding capacity (TIBC) were normal. As for the total IgE dosage, it was elevated at 68 UI/mL with class 2 allergy to Hazelnuts and class I allergy to Wheat flour.

The diarrhea persisted, was getting worse in frequency and became bloody with mucus. So at the age of 10 months, a full gastrointestinal endoscopy was done. It showed active eosinophilic duodenitis with elevated intraepithelial lymphocytes (LIE), atrophic chronic gastritis with eosinophils, eosinophilic esophagitis, gastritis and eosinophilic colitis. Pathology report confirmed an eosinophilic infiltration limited to the mucosal layer with rare eosinophilic abscesses of the esophagus, stomach and colon.

3. DISCUSSION

Our patient presentation is similar to any patient with neonatal sepsis. The peculiar features are the persistently abnormal CBC -namely the elevated WBC count and eosinophils- with an elevated CRP. Aside from the gastrointestinal manifestations that started around 2 ½ months of age, the patient was clinically well. He was growing on the third percentile for weight up till 5 months of age when he dropped from his growth curve for weight.

Family history was negative for any similar problems and was positive only for asthma in his older brother.

GI endoscopy along with the pathology report confirmed the diagnosis of Eosinophilic Gastroenteritis.

Eosinophilic Gastroenteritis is a triad of disturbed GI function with elevated eosinophils on peripheral smear and eosinophilic infiltration of the gastrointestinal tract .Other causes of eosinophilia should be excluded like sepsis, anabolic state, drug reactions, response to foreign antigens, chronic lung disease and erythropoietin treatment [3]. Eosinophilia is highly associated with sepsis in the newborn [4].

The persistence of eosinophilia beyond the neonatal period with worsening GI symptoms as diarrhea, vomiting, failure to thrive and intolerance to any solid food in spite of the patient being on an elemental formula should be highly suggestive of eosinophilic gastroenteritis. An upper and lower GI endoscopy should be ordered to confirm this diagnosis since it may affect any part of the GI tract hence the heterogeneity in its presentation.

Most patients with eosinophilic gastroenteritis respond to conservative measures, empiric dietary elimination [5] and oral corticosteroids [6]. Patients can be monitored every 6 months. Biologic therapy with humanized interleukin 5 (IL-5) treatment has been associated with a reduced eosinophil count but not with improved clinical symptoms [7]. Treatment with omalizumab has shown a reduction in eosinophil count as well as clinical improvement of symptoms; however, the results were not clinically significant [8]. A 2016 case report showed treatment with clarithromycin resulted in both improvement in clinical symptoms and a decrease in eosinophil count [9].

Our patient received an oral dose of prednisolone 2 mg/kg for 1 month which was tapered over 2 months. This resulted in improvement of her blood count: WBC 13900/ul, Neutrophils 33%, Eos 1% and CRP 6 mg/l. The growth chart showed catch up in weight as shown in the growth chart. Montelukast was also started after 2 months of steroids [10].

4. CONCLUSION

This is a challenging case of eosinophilic gastroenteritis in a newborn that presented at 3 days of life. The literature review shows that the average age of diagnosis is 8.3 years [10]. Although rare, awareness about this disorder along with a high index of suspecting it should be present in any newborn with persistently elevated eosinophils and worsening GI symptoms after excluding common causes for neonatal eosinophilia. A Multidisciplinary team approach is essential for early diagnosis and management of this entity.

CONSENT

As per international standard or university standard, patient's written consent has been collected and preserved by the authors.

ETHICAL APPROVAL

As per international standard or university standard, written approval of Ethics committee has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Ingle SB HIC. Eosinophilic gastroenteritis: An unusual type of gastroenteritis. World J Gastroenterol. 2013;19:5061-5066.
2. Nancy Gupta, Arun Aggarwal RG. The management of eosinophilic gastroenteritis. Scand J Gastroenterol. 2015;50:1309-1314.
3. Manoura A, Hatzidaki E, Korakaki E, Mitsaki M, Valari V GC. Eosinophilia in sick neonates. Hematol. 2002;32(1):31-37.
4. Jagtap SV, Nikumbh DB, Kshirsagar A. Unusual presentation of eosinophilic enteritis as multiple strictures of small intestine. Clin Pract. 2012;3(1).
5. Lucendo AJ, Serrano-Montalbán B, Arias Á, Redondo O, TJ. Efficacy of dietary treatment for inducing disease remission in eosinophilic gastroenteritis. J Pediatr Gastroenterol Nutr. 2015;61(1):56-64.
6. Cianferoni A, Spergel JM. Eosinophilic esophagitis and gastroenteritis. Curr Allergy Asthma Rep. 2015;15(9):58.
7. Kim YJ, Prussin C, Martin B, Law MA, Haverty TP, Nutman TB KA. No TitleRebound eosinophilia after treatment of hypereosinophilic syndrome and eosinophilic gastroenteritis with monoclonal anti-IL-5 antibody SCH55700. J Allergy Clin Immunol. 2004;114(6):1449-1455.
8. Foroughi S, Foster B, Kim N, Bernardino LB, Scott LM, Hamilton RG, Metcalfe DD, Mannon PJ, Calman Prussin. Anti-IgE treatment of eosinophil-associated gastrointestinal disorders. J Allergy Clin Immunol. 2007;120(3):594-601.
9. Naw April Phaw HHT. Eosinophilic gastroenteritis: A challenge to diagnose and treat. BMJ Case Reports 2016. 2016;10.
10. Tien F, Wu J, Jeng Y, et al. Clinical Features and treatment responses of children with eosinophilic gastroenteritis. Pediatr Neonatol. 2011;52(5):272-278. DOI: 10.1016/j.pedneo.2011.06.006

Peer-review history:
The peer review history for this paper can be accessed here:
http://sciencedomain.org/review-history/20569