

Henry Ford Health System

Henry Ford Health System Scholarly Commons

Case Reports

Medical Education Research Forum 2020

5-2020

Persistent hypogammaglobulinemia in CVID secondary to protein losing enteropathy

Sunjay Modi

Henry Ford Health System, smodi4@hfhs.org

Alex Horbal

Henry Ford Health System, ahorbal1@hfhs.org

Jasmine F. Alsukhon

Henry Ford Health System, jalsukh1@hfhs.org

Haejin Kim

Henry Ford Health System, hkim3@hfhs.org

Follow this and additional works at: <https://scholarlycommons.henryford.com/merf2020caserpt>

Recommended Citation

Modi, Sunjay; Horbal, Alex; Alsukhon, Jasmine F.; and Kim, Haejin, "Persistent hypogammaglobulinemia in CVID secondary to protein losing enteropathy" (2020). *Case Reports*. 48.

<https://scholarlycommons.henryford.com/merf2020caserpt/48>

This Poster is brought to you for free and open access by the Medical Education Research Forum 2020 at Henry Ford Health System Scholarly Commons. It has been accepted for inclusion in Case Reports by an authorized administrator of Henry Ford Health System Scholarly Commons.



Introduction

- We report a case of a patient with Common Variable Immunodeficiency (CVID) presenting with septic shock and persistent hypogammaglobulinemia despite adequate replacement due to protein losing enteropathy and chronic wounds causing protein loss.
- CVID should be suspected in individuals with reduced levels of serum IgG in combination with low levels of IgA and/or IgM, reduced response to immunizations and an absence of any other immunodeficiency state [1]. Figure 1 demonstrates pathogenesis of CVID.
- CVID is treated with monthly IVIG infusions, which is the treatment regimen our patient was receiving.

Case Presentation

- Patient is a 40 year old male with PMH of B cell Lymphoma s/p R-CHOP and radiation in remission, Common Variable Immunodeficiency (CVID) diagnosed prior to the hospital admission after IgG levels were found to be <75 g/dL (reference range of 700-1600 mg/dL) and recurrent *Clostridium difficile* (*C. difficile*) colitis admitted to the ICU for left hip abscess and *Acinetobacter* pneumonia causing septic shock requiring ICU admission.
- Patient had a history of non-healing joint infections requiring multiple antibiotics and surgeries leading to persistent wounds causing serosanguinous drainage.
- Patient was found to have IgG level was 142 mg/dL (reference range of 700-1600 mg/dL) upon admission and he received 30g of IVIG.
- One day post-IVIG infusion, IgG levels were 565 mg/dL.
- Fourteen days after the infusion his IgG dropped down to 272 mg/dL.
- There was high suspicion patient had a disease process leading to acute protein loss, UA did not demonstrate nephrotic syndrome, therefore it was likely patient had protein losing enteropathy from recurrent *C. difficile* colitis which lead to persistent diarrhea.
- Patient's hypogammaglobulinemia persisted due to chronic wounds from surgical sites causing protein loss (Figure 2) superimposed with protein losing enteropathy.
- Due to overwhelming multifocal infections, per family wishes, patient was transitioned to comfort care and passed away peacefully.

Diagrams and Clinical Imaging

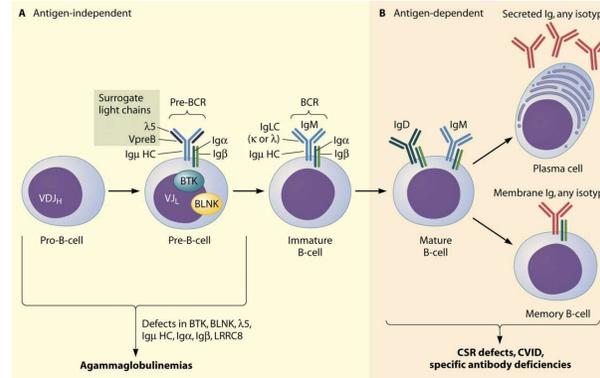


Figure 1: Pathogenesis of CVID [6]



Figure 2: Left hip s/p surgical intervention with serosanguinous drainage

Discussion

- We present a case of a patient with known history of CVID who presented with septic shock and lack of normal IgG levels after recent IVIG infusion due to protein losing enteropathy and protein loss from chronic wounds.
- Our patient was tested for an immunoglobulin deficiency prior to hospital admission as patient had multiple courses of failure of antibiotics for joint infections.
- This individual was diagnosed with CVID in 2019 as IgG<75 mg/dL (L) with concurrent IgA <10 mg/dL (L) and IgM< 20 mg/dL (L). It was thought the immunoglobulin deficiency was a secondary hypogammaglobulinemia due to Rituximab [2], but there was strong clinical suspicion for true CVID given the severity of his infections.

- When this patient's persistent hypogammaglobulinemia was discovered even after IgG infusions, reversible causes were looked into such as nephrotic syndrome.
- There have been notable cases of protein losing enteropathies, such as Celiac Disease causing secondary hypogammaglobulinemia or exacerbating the primary disease process, such as CVID [3].
- It is likely that patient's acute drop in IgG 14 days after IgG infusions was due to protein losing enteropathy secondary to recurrent *C. difficile* diarrhea and protein loss from significant serosanguinous drainage from chronic wounds [4,5].

Conclusions

- Secondary hypogammaglobulinemia may occur after Rituximab therapy.
- This case presentation demonstrates the utility in checking IgG levels prior to Rituximab therapy as there may be worsening of immunoglobulin levels post-Rituximab therapy.
- Acute protein loss can exacerbate CVID and cause overwhelming infections leading to septic shock.
- It is important to determine the etiology of acute protein loss early on in the disease process with underlying CVID as reversible causes that are identified may improve patient outcomes or higher dosing of IVIG may be indicated in such settings where there is a rapid decline of immunoglobulin.

Bibliography

- Hammarström L, Vorechovsky I, Webster D. Selective IgA deficiency (SIgAD) and common variable immunodeficiency (CVID). *Clin Exp Immunol*. 2000;120(2):225–231. DOI:10.1046/j.1365-2249.2000.01131.x
- Mehdi Mohammad, G. Adeli, Brian H. Eichner, Courtney Thornburg & Larry Williams. Persistent Antibody depletion after Rituximab in three children with autoimmune cytopenias. *Pediatric Hematology and Oncology*. 2009.26:8, 566-572, DOI: 10.3109/08880010903271697
- Ameratunga, R., Barker, R.W., Steele, R.H. *et al*. Profound Reversible Hypogammaglobulinemia Caused by Celiac Disease in the Absence of Protein Losing Enteropathy. *J Clin Immunol*. 2015. **35**, 589–594. DOI: 10.1007/s10875-015-0189-6
- Radet C, Ginies JL, Coirier F, Person B, Meunier M, Limal JM. A rare cause of severe diarrhea in children: pseudomembranous colitis. *Arch Pediatr*. 1995 Nov;2(11):1081 5. DOI:10.1016/0929-693x(96)81284-x
- Lizaka S, Sanada H, Nakagami G, Sekine R, Koyanagi H, Konya C, Sugama J. Estimation of protein loss from wound fluid in older patients with severe pressure ulcers. *Nutrition*. 2010. Sep;26(9):890-5. doi: 10.1016/j.nut.2009.09.008
- Fried A, Francisco B. Pathogenesis, Diagnosis, and Management of Primary Antibody Deficiencies and Infections. *Clinical Microbiology Reviews*. 2009. July. 22 (3) 396-414; DOI: 10.1128/CMR.00001-09