

Case Study

**LOW CD 4 COUNT IN HIV NEGATIVE PATIENT-A CASE OF COMMON VARIABLE DEFICIENCY
CENTRAL HOSPITAL NWR JAIPUR DEPT OF MEDICINE: A CASE REPORT**

RAKESH KUMAR MEENA*, DEEPAK AGARWAL, MANISH PABRI

Central Railway Hospital, Jaipur, Rajasthan, India

*Corresponding author: Rakesh Kumar Meena; *Email: Jaipuraarkeyem4u@gmail.com

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ABSTRACT

Common variable immunodeficiency syndrome (CVID) is a disorder characterised by reduced levels of IgG, IgA and/or IgM, and recurrent bacterial infections. Sinopulmonary infections, gastrointestinal infections. It is commonly seen in association to immunodeficiency states such as CVID, IgA deficiency and chronic infections due to Giardia lamblia and Helicobacter pylori. We report a case of CVID with low CD 4 counts having recurrent diarrhoea, weight loss, anasarca and Hypoproteinaemia.

Keywords: CVID-common variable immunodeficiency, Recurrent diarrhea and hypoproteinemia, HIV-human immunodeficiency virus, CD 4-cluster of differentiation

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INTRODUCTION

Common variable immunodeficiency (CVID) is a condition of low serum level of one or more immunoglobulin [1]. The condition is recognized predominantly in adults, although clinical manifestations can occur in early life. Hypogammaglobulinemia is associated with at least partially defective antibody production in response to vaccine antigen. B lymphocyte count is often normal but can be low. Diagnostic criteria are: -recurrent infections; -age more 4 y; -reduced levels of IgG; decrease of IgA and/or IgM; exclusion of the other causes of hypogammaglobulinemia; -no isohemagglutination and response to the vaccine. Five clinical phenotypes are described: only infections, autoimmunity, polyclonal lymphocytic infiltration, malignancy and enteropathy. The treatment of patients with CVID is performed with the use of an infusion of immunoglobulin [2]. The incidence of CVID is estimated to be between 1:25 000 and 1:50 000 in white individuals, but it is apparently less common in Asian and African American individuals. It is equally prevalent in males and females. In all studies of CVID, a delay in making the diagnosis has been noted [3]. Sinopulmonary infections are most commonly reported followed by gastrointestinal infections [3]. Persistent diarrhoea malabsorption and Liver disease is common Gastrointestinal manifestation. CVID subjects had lower numbers of CD3+CD4+T cells and higher numbers of CD3+CD8+T cells and IgG deficient patients [4].

Case history and evaluation

A 39 y old age male case presented to us with history of recurrent diarrhea-4 y, weight loss and anasarca without liver cirrhosis, renal illness and cardiac disorder. Patient is nonalcoholic, No history of similar

illness in family and consanguinity. On routine examination and clinical evaluation, he was found suffering from edema feet with postural hypotension. Hemoglobin-14.40 g/dl, Total Leucocyte count-7170/cumm, Neutrophil-72%, Lymphocytes-21.30%, Eosinophil-2.06%, Plateles-1.9 lac/mm³. ESR-10 mm/in 1st h. Total Protein-4.0 gm% (hypoproteinemia), S. Albumin-2.6 gm% and Serum globulins were very low 0.96 mg/dl while SGOT-44 U/l, SGPT-53 U/l, S. Bilirubin-0.3 mg/dl. PT/INR-13 sec/1.23. CRP-3.78 mg/l. Renal Function test was normal, Serum Electrolytes were normal, 24 H Urinary Protein-Normal, Albumin creatinine ratio-27.7 mg/g-Normal. HIV-Negative. TSH-4.11 mcIU/ml. Echocardiography was normal with LVEF-60% and All Valve were normal; ASCA-Negative, ANA-Negative, and Alpha1anti-trypsin levels were-not reduced. Tissue transglutaminase-IgA-Negative.

CRP-3.78 mg/l. Chest Xray-Normal and no thymus shadow, No cardiomegaly and Normal lung Parenchyma. Stool for Ova and cyst examination remain negative. Tissue transglutaminase antibody-<3 AU/ml s/o negative, and USG Abdomen s/o Mild Hepatomegaly.

As the reports suggestive of both low albumin and low globulin so Serum immunoglobulins and CD 4 count and Lymphangiography was done in view of possible illness like CVID, Lymphangiectasia. His G. I. evaluation was done in view of Protein-losing enteropathy and Duodenal Biopsy was suggestive of nonspecific duodenitis with normal intraepithelial lymphocytes, crypt: villous ratio was 1:3 to 1:2, cryptic hyperplasia was present with mild inflammation of Lamina propria, no dilated lacteals were present and H. pylori-associated gastritis. Video capsule endoscopy-Normal, MRI Lymphangiography was done in view of lymphangiectasia but did not show any dilated lymphatics and any area of extravasation.

Table 1: Comprehensive diagnostic evaluation and management of A 39 y old male with common variable immunodeficiency (Cvid)

Immunoglobulin	Measured value	Normal range	Interpretation
S. IgM	30.9 mg/dl	24-230 mg/dl	Low
S. IgA	33.9 mg/dl	70-400 mg/dl	Low
S. IgG	267 mg/dl	700-1600 mg/dl	Low
S. IgE	5 mg/dl	2-214 mg/dl	Normal

TLC-5360 Cells/cumm, lymphocyte-14.70%, CD 3 Cells-63% (60-90% normal), CD4 Counts-166 Cells/mm³-21% OF CD3 of total TLC-5360 CELLS/CUMM (normal range is 30-50 OF CD3 by flow cytometry and high CD8 count-41% (10-31% NORMAL) percentage among lymphocytes.

On the basis of all investigations and clinical presentation patient was diagnosed as a case of Common Variable Immunodeficiency

(CVID) He was started Iv Ig infusion every 3 to 4 wly and patient got symptomatically improved; his frequent episode of diarrhea

stopped, patient also gained weight. Patient was redeployed from a train-LoCo Pilot public safety job to an alternate desk job on permanent basis. He is in regular follow-up and better.

DISCUSSION

This patient is found suffering from long-duration recurrent diarrhea and used to respond to antibiotics when he started developing weight loss and anasarca. He was found suffering from hypoproteinemia and low globulin levels, while his renal function and cardiac functions was normal. On further evaluation, he was diagnosed with low immunoglobulins, making him prone for recurrent acute diarrhea episodes as his chest x-ray, Video capsule endoscopy and Lymphangiography was normal. It was an uncommon presentation so his flow cytometric analysis and serum immunoglobulin assay was done as reduced CD4 count with Negative HIV status, reduced IgG, IgA, IgM with Normal IgE and High CD 8 % of Lymphocytes was pointing toward the diagnosis of CVID. Recurrent diarrhea is one of the common presentation of CVID so we should keep high index of suspicion for immunodeficiency syndrome like CVID as there is a different type treatment for such illnesses [5].

Differential diagnosis

1. Good's syndrome, an infrequent adult-onset immunodeficiency, is characterized by the triad of thymoma, hypogammaglobulinemia, and increased susceptibility to recurrent infections in our case there is no thymoma in x ray chest [6].
2. Protein losing enteropathy-Patient who have only Protein losing enteropathy did not develop recurrent infections and have normal IgA levels, while in our patient IgA levels were low and he also had history of recurrent infections [7].
3. HIV Infection-As Patient was negative for HIV testing several times.
4. Hyper IgM Syndrome-As CD154+also known as CD40 L which was found normal [8].

Learning Point-Among healthcare worker's awareness should be increased to suspect such type of illness like CVID in patients suffering from long duration frequent diarrhoea multiple episodes.

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AUTHORS CONTRIBUTIONS

All the authors have contributed equally.

CONFLICTS OF INTERESTS

Declared none

REFERENCES

1. Fischer A. Primary immune deficiency diseases. In: Loscalzo J, Fauci A, Kasper D, Hauser S, Longo D, Jameson JL. editors. Harrison's principles of internal medicine. 21st ed. New York: McGraw-Hill Education; 2022. Available from: <https://accessmedicine.mhmedical.com/content.%20aspx?aid=1190522036>
2. Moore D, Dias F, Esberard E, Mugayar J, Costa M, Pestana S. Common variable immunodeficiency misdiagnosed as crohn disease. *World Allergy Organ J.* 2015;8:A267. doi: 10.1186/1939-4551-8-S1-A267.
3. Cunningham-Rundles C. Common variable immune deficiency: case studies. *Hematology Am Soc Hematol Educ Program.* 2019;(1):449-56. doi: 10.1182/hematology.2019002062, PMID 31808912.
4. Daza Cajigal V, Segura Guerrero M, Lopez Cueto M, Robles Marhuenda A, Camara C, Gerra Galan T. Clinical manifestations and approach to the management of patients with common variable immunodeficiency and liver disease. *Front Immunol.* 2023;14:1197361. doi: 10.3389/fimmu.2023.1197361, PMID 37342345. Available from: <https://www.frontiersin.org/journals/immunology/articles/10.3389/fimmu.2023.1197361>
5. Franzblau LE, Fuleihan RL, Cunningham Rundles C, Wysocki CA. CVID-associated intestinal disorders in the USIDNET registry: an analysis of disease manifestations, functional status, comorbidities, and treatment. *J Clin Immunol.* 2023;44(1):32. doi: 10.1007/s10875-023-01604-6, PMID 38133694.
6. Tamburello A, Castelnovo L, Faggioli P, Bompane D, Brando B, Gatti A. Good's syndrome, a rare form of acquired immunodeficiency associated with thymomas. *Clin Pract.* 2019;9(2)/2039:1112. doi: 10.4081/cp.2019.1112, PMID 31240091.
7. Sanges S, Germain N, Vignes S, Seguy D, Stabler S, Etienne N. Protein-losing enteropathy as a complication and/or differential diagnosis of common variable immunodeficiency. *J Clin Immunol.* 2022 Oct;42(7):1461-72. doi: 10.1007/s10875-022-01299-1, PMID 35737255.
8. Ochs HD, Hollenbaugh D, Aruffo A. The role of CD40L (gp39)/CD40 in T/B cell interaction and primary immunodeficiency. *Semin Immunol.* 1994;6(5):337-41. doi: 10.1006/smim.1994.1042, PMID 7532462.