

A Case Report and Review of Literature: A Patient With Common Variable Immunodeficiency and Pericardial Effusion



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ABSTRACT

Introduction: Common Variable Immunodeficiency (CVID) is one of the primary immunodeficiencies that its patients can develop its symptoms since infancy till senility; however, it usually manifests between the age of 15 and 40 years. It is characterized by low antibody levels and recurrent infections. Individuals with CVID are more prone to autoimmune diseases and malignancy.

Case presentation: We presented a 17-year-old girl with a documented CVID, who was treated with intravenous immunoglobulin and prophylactic antibiotics. She suffered from CVID from the age of 13 to the study time. She was hospitalized due to illness, fever, and severe dyspnea. Her chest X-ray revealed cardiomegaly; thus, high-resolution thorax CT scan and echocardiography were performed which revealed pericardial effusion. No underlying autoimmune diseases were detected in our assessments. Significant clinical, radiographic and physiological improvements were achieved after conducting an appropriate therapy. The patient was followed for two years and showed no clinical and laboratory findings in respect to autoimmune diseases. In our case, the pericardial effusion was present without any evidence of autoimmunity.

Conclusions: Cardiac manifestation and complications are rare in CVID patients in the absence of an autoimmune disease; however, they should be considered.

1. Introduction

Common Variable Immunodeficiency (CVID) is among severe antibody deficiencies. It is one of the most frequent primary immune deficiencies in adults with an estimated prevalence of 1 per 25000-100000. It is the most common symptomatic primary immunodeficiency

after isolated Immunoglobulin A (IgA) deficiency. This disease is caused by immune deregulations resulting in failed B-cell differentiation with reduced immunoglobulins production. It is sometimes diagnosed in childhood; however, it typically occurs after puberty. Nevertheless, there could be a delay in its diagnosis. The majority of patients are diagnosed between the ages of 20 and 45 years (1-5).

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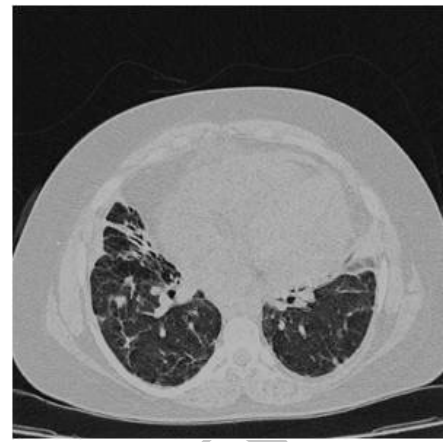
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Figure 1. Chest X-ray of the patient

CVID is characterized by hypogammaglobulinemia, poor to absent specific antibody responses to vaccination, increased susceptibility to infections and some complications such as splenomegaly, autoimmunity, lymphoproliferation, malignancy, and granulomas (6-8). Bronchiectasis occurs in 17-76% of cases, and cardiovascular complications such as pulmonary hypertension and heart failure rarely occur (9). Thus, many organs are involved in CVID. However, the cardiac symptoms like pericardial effusion is rare in the absence of autoimmune diseases (e.g. systemic lupus erythematosus). Therefore, we aimed to discuss an interesting case of CVID complicated with pericardial effusion without any autoimmune underlying diseases.

2. Case Presentation

A 17-year-old Iranian girl was admitted with a history of recurrent sinopulmonary infections and several hospitalizations since the age of 5 years. She was referred to many physicians, and immunodeficiency and hypo-



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Figure 2. Lung HRCT scan

gammaglobulinemia examinations revealed her poor responses to vaccines. Due to these findings, flow cytometry analysis was performed in her at the age of 14 that was in favor of CVID. The main treatment consisted of Intravenous Immunoglobulin (IVIg) infusion during previous years and prophylactic antibiotics use.

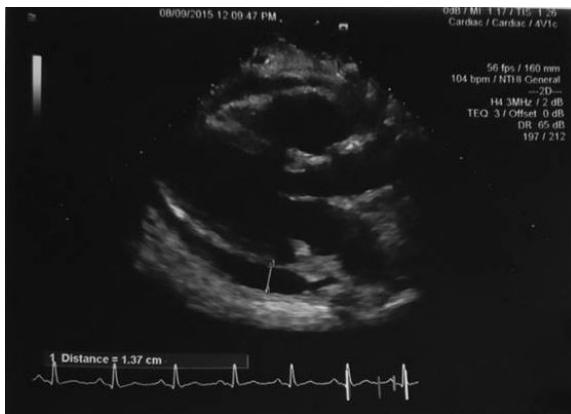
Despite receiving IVIg and prophylactic antibiotics, she suffered from fever and severe dyspnea and was admitted to the emergency room. Her Chest X-Ray (CXR) revealed a linear and alveolar infiltration pattern in the right lower lobe of the lung, a massive consolidation, and cardiomegaly (Figure 1). A high-resolution thorax CT scan was performed without a contrast, which was significant for ground glass opacities and intralobular septal thickening with the lower lobe predominance and pericardial effusion (Figure 2).

Echocardiography demonstrated moderate to large pericardial effusion mostly in the posterior and lateral sides of LV with EF: 60% (Figure 3). In the pathological

Table 1. The list of some publications on cardiac complications

Author(s)	Publication Year	Complication(s)	Patient(s)
Laufs et al. (17)	2002	Giant cell myocarditis	A 12-year-old girl
Kashef et al. (16)	2011	Giant aneurysm of the thoracic and proximal abdominal aorta	A 16-year-old boy
Jerschow et al. (18)	2007	Takayasu arteritis	A 53-year-old Hispanic woman
Dave et al. (15)	2007	Myocardial infarction with IVIg infusion	A 65-year-old man
Johnston et al. (19)	2004	Aortic root dilatation	
Yalchin et al. (20)	2004	Asymptomatic right-sided aortic arch	A 41-year-old woman
Cambrey Gutierrez et al. (9)	2015	Valvular insufficiency and pulmonary hypertension	

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Figure 3. Patient's echocardiogram revealing pericardial effusion

microscopic assessment of pericardial effusion, numerous inflammatory cells were observed, and polymorphonuclear were the most frequent leukocytes. However, no evidence of malignancy was detected. The pericardial fluid analysis findings were in favor of transudative pericardial effusion. The patient recovered without any surgical procedure like drainage tube placement or chest tube insertion. Immunologic work-up for R/O autoimmune diseases (e.g. SLE) such as antinuclear antibody, anti-double strand DNA, P&C Anti-neutrophil cytoplasmic antibodies, and anti-cardiolipin were performed with negative results. The patient was followed for two years and showed no clinical and laboratory findings indicating autoimmune diseases.

3. Discussion

The clinical manifestations of CVID are multifarious; among which, recurrent bacterial infections caused by encapsulated bacteria are the most common ones. These infections lead to sinusitis, otitis media, bronchitis, and pneumonia. In untreated patients, chronic sinusitis and bronchiectasis are frequent complications leading to significant morbidity and mortality (10-12). Autoimmunity occurs in approximately 25% of the patients with CVID (13). The most common autoimmunity is cytopenia. However, other autoimmune patients such as thrombocytopenic purpura, hemolytic anemia or neutropenia are also at higher risks. Pericardial effusion is rare in CVID; however, some viruses (CMV, HIV, coxsackieviruses, and echoviruses), drugs (isoniazid, hydralazine, and phenytoin), and autoimmune diseases (SLE in particular) can cause pericardial effusion.

4. Review of Literature

Cardiac involvement is not prevalent in CVID. However, pericardial effusion can be present in autoimmunity dis-

eases like SLE (14). Other cardiac complications can be secondary to bronchiectases like pulmonary hypertension and heart failure. Myocardial infarction, myocarditis, heart failure, pulmonary arteries hypertension, and giant aneurysm of aorta were reported among cardiac problems in antibody deficiencies; however, pericardial effusion was never observed (15, 16). Some of these complications are listed in Table 1.

Cardiac manifestation and complications are not very common in CVID patients in the absence of an autoimmune disease. In our studied case, pericardial effusion was present without any evidence of autoimmunity or drug induction. Although a pericardial effusion is rare, it should be considered, and complete autoimmune disease assessments should be performed in the follow-up period.

Ethical Considerations

Compliance with ethical guidelines

Informed consent was obtained from the human subject of this study.

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Authors contributions

All authors contributed in preparing this article.

Conflict of interest

The authors declared no conflict of interest.

References

- Gathmann B, Mahlaoui N, Gerard L, Oksenhendler E, Warnatz K, Schulze I, et al. Clinical picture and treatment of 2212 patients with common variable immunodeficiency. *The Journal of Allergy and Clinical Immunology*. 2014; 134(1):116-26. [DOI:10.1016/j.jaci.2013.12.1077] [PMID]
- Paranjpe SM, Koticha A, Mehta PR. Chronic giardiasis in a case of Common Variable Immunodeficiency (CVID): A case report. *Journal of Clinical and Diagnostic Research*. 2016; 10(7):DD03-DD04. [DOI:10.7860/JCDR/2016/16138.8183]

3. Rezaei N, Abolhassani H, Kasraian A, Mohammadinejad P, Sadeghi B, Aghamohammadi A. Family study of pediatric patients with primary antibody deficiencies. *Iranian Journal of Allergy, Asthma and Immunology*. 2013; 12(4):377-82. [PMID]
4. Yalavarthy UC, Panda M. A middle-aged female with recurrent sinopulmonary infections: A case report. *Journal of Medical Case Reports*. 2008; 2(1):117. [DOI:10.1186/1752-1947-2-117] [PMID] [PMCID]
5. Aghamohammadi A, Abolhassani H, Latif A, Tabassomi F, Shokuhfar T, Torabi Sagvand B, et al. Long-term evaluation of a historical cohort of Iranian common variable immunodeficiency patients. *Expert Review of Clinical Immunology*. 2014; 10(10):1405-17. [DOI:10.1586/1744666X.2014.958469] [PMID]
6. Abbott JK, Gelfand EW. Common variable immunodeficiency: Diagnosis, management, and treatment. *Immunology and Allergy Clinics of North America*. 2015; 35(4):637-58. [DOI:10.1016/j.iac.2015.07.009] [PMID]
7. Resnick ES, Cunningham-Rundles C. The many faces of the clinical picture of common variable immune deficiency. *Current Opinion in Allergy and Clinical Immunology*. 2012; 12(6):595-601. [DOI:10.1097/ACI.0b013e32835914b9] [PMID]
8. Saikia B, Gupta S. Common variable immunodeficiency. *Indian Journal of Pediatrics*. 2016; 83(4):338-44. [DOI:10.1007/s12098-016-2038-x] [PMID]
9. Cambray-Gutiérrez JC, Fernández-Muñoz MJ, Del Rivero-Hernández LG, López-Pérez P, Chávez-García AA, Segura-Méndez NH. Structural and functional heart diseases in adult patients with common variable immunodeficiency. *Revista Alergia México*. 2015; 62(2):91-7.
10. Lougaris V, Tabellini G, Vitali M, Baronio M, Patrizi O, Tampella G, et al. Defective natural killer-cell cytotoxic activity in NFKB2-mutated CVID-like disease. *The Journal of Allergy and Clinical Immunology*. 2015; 135(6):1641-3. [DOI:10.1016/j.jaci.2014.11.038] [PMID]
11. Ochs HD. Common Variable Immunodeficiency (CVID): New genetic insight and unanswered questions. *Clinical and Experimental Immunology*. 2014; 178(S1):5-6. [DOI:10.1111/cei.12491] [PMID] [PMCID]
12. Paquin-Proulx D, Sandberg JK. Persistent immune activation in CVID and the role of IVIg in its suppression. *Frontiers in Immunology*. 2014; 5:637. [DOI:10.3389/fimmu.2014.00637] [PMID] [PMCID]
13. Boileau J, Mouillot G, Gerard L, Carmagnat M, Rabian C, Oksenhendler E, et al. Autoimmunity in common variable immunodeficiency: Correlation with lymphocyte phenotype in the French DEFI study. *Journal of Autoimmunity*. 2011; 36(1):25-32. [DOI:10.1016/j.jaut.2010.10.002] [PMID]
14. Frieri M. Good's syndrome, CVID, and selective antibody deficiency in patients with chronic rhinosinusitis. *Current Allergy and Asthma Reports*. 2014; 14(6):438. [DOI:10.1007/s11882-014-0438-4] [PMID]
15. Dave S, Hagan J. Myocardial infarction during intravenous immunoglobulin infusion in a 65-year-old man with common variable immunodeficiency and subsequent successful repeated administration. *Annals of Allergy, Asthma & Immunology*. 2007; 99(6):567-70. [DOI:10.1016/S1081-1206(10)60388-2]
16. Kashef S, Moghtaderi M, Borzouee M, Rezaei N. Giant aneurysm of thoracic and proximal abdominal aorta in a patient with common variable immunodeficiency. *Iranian Journal of Allergy, Asthma and Immunology*. 2011; 10(2):133-7. [PMID]
17. Laufs H, Nigrovic PA, Schneider LC, Oettgen H, del Nido P, Moskowitz IPG, et al. Giant cell myocarditis in a 12-year-old girl with common variable immunodeficiency. *Mayo Clinic Proceedings*. 2002; 77(1):92-6. [DOI:10.4065/77.1.92] [PMID]
18. Jerschow E, De Vos GS, Hudes G, Rubinstein A, Lipsitz EC, Rosenstreich D. A case of common variable immunodeficiency syndrome associated with Takayasu arteritis. *Annals of Allergy, Asthma & Immunology*. 2007; 98(2):196-9. [DOI:10.1016/S1081-1206(10)60697-7]
19. Johnston S, Hill S, Lock R, Dwight J, Unsworth D, Gompels M. Echocardiographic abnormalities in primary antibody deficiency. *Postgraduate Medical Journal*. 2004; 80(942):214-8. [DOI:10.1136/pgmj.2003.012443] [PMID] [PMCID]
20. Yalcin AD, Aydemir N, Turgut H, Erbay RH, Yalcin AN. Common variable immunodeficiency syndrome with right aortic arch: A case report. *BMC Infectious Diseases*. 2004; 4:2. [DOI:10.1186/1471-2334-4-2] [PMID] [PMCID]