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Case Report

Disseminated bronchiectasis in an adult with common variable immunodeficiency

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**Introduction**

Common Variable Immunodeficiency (CVID) is a predominantly antibody primary immunodeficiency in which the humoral immune response is altered. The clinical spectrum of this disease ranges from repeated infections with sequelae such as the appearance of bronchiectasis, to the development of malignancies or autoimmunity. Despite being a genetic disorder, adults are the most affected, so efforts should be attempted to educate medical community. Here we present the case of a 37-year-old man with recurrent sinopulmonary infections and widespread bronchiectasis, in whom a severe hypogammaglobulinemia with symptoms compatible with Common Variable Immunodeficiency was demonstrated.

**Case description**

A 37-year-old man presented to the emergency department of a level III hospital in the city of Cali (Colombia) complaining of respiratory distress, fever and cough with greenish expectoration of approximately one week duration, with worsening dyspnea in the past 48 hours until being unable of performing any minimal effort. At admission, he presented hypotension (78/36), tachycardia (126 beats/min), and tachypnea (38 breaths /min), with saturation of 76% O₂ (O₂ atmosphere); lung auscultation revealed multiple over-aggregate and overall decreased breath sounds. The patient reported having immunodeficiency antibody. Few minutes after admission, he presented respiratory failure requiring intubation and vasoactive support with mechanical ventilation. On suspicion of septic shock, antibiotic coverage was initiated with vancomycin and cefepime, after taking blood cultures.

The patient is native to, and came from Cali (Valle province, in Colombia). As relevant background, he refers pneumonia, sinusitis and recurrent otitis since he was aged 18 yrs, with countless episodes (6 to 10 per year) requiring long courses of oral or intravenous antibiotics and multiple hospitalizations. Since 2002 cylindrical and cystic bronchiectasis had been documented in all four quadrants (Fig. 1A), equally documented in the cross sections at the level of the aortic arch and the left ventricle (Fig. 1B y1C).

![Figure 1. High resolution chest scans which show widespread bronchiectasis in the four quadrants (A); and in the cross sections at the level of the aortic arch (B), and the left ventricle (C).](image-url)
Primary immunodeficiencies (PID) are considered rare diseases (prevalence 1: 25,000 to 1: 50,000), which results in a significant delay in diagnosis that leads to impoverished prognosis, increased comorbidities and clinical worsening, and increased cost to the health system for not being this condition timely identified. Traditionally, it has been accepted that PID are diseases of childhood, so internal medicine rarely addresses them, which contributes to the fact that adult doctors are not familiar with this disorder.

Common Variable Immunodeficiency (CVID) is the primary immunodeficiency of clinical relevance most frequently found in adulthood. Traditionally, it is accepted that this disease has two peaks of incidence, the former in childhood and the latter between the second and third decades of life, as it is the case of our patient. The delay in diagnosis is common, occurring 2, 15 or even 20 yrs after the onset of symptoms.

Common Variable Immunodeficiency is a very heterogeneous disease in which numerous mutations associated with maturation or activation of B lymphocytes have been identified, which ultimately results in both a quantitative and qualitative inability to produce immunoglobulin genes. Today, two main clinical spectra are recognized: uncomplicated CVID, referred to the “classical” patient with repetitive sinopulmonary infections or gastrointestinal complications; and complicated CVID, in which recurrent infections occur, accompanied by visceromegalies (hepato/splenomegaly), lymphadenopathy, autoimmunity (usually cytopenias), and lymphomas. The presence of widespread bronchiectasis, as in the case of our patient, requires an active seeking and ruling out of primary immunodeficiencies, as it has been shown that up to 10% of patients with bronchiectasis (not caused by cystic fibrosis) correspond to antibody deficiencies, especially CVID.

The diagnostic criteria for CVID were established by the European and Panamerican Immunodeficiencies Societies (ESID/PAGID) in 1999, and still in force so far, include: marked decrease (at least 2 standard deviations below the mean for age) of IgG, IgA and/or IgM in serum; to be older than 4 years, to be negative for isohemagglutinins and/or to have poor responses to vaccines; besides, other causes of hypogammaglobulinemia must have been excluded. Compliance with all of the above criteria is essential for diagnosis prior to initiation of therapy with IVIG replacement, because this therapy modifies serological parameters up to six months after the last application.

The presence of lymphopenia at the expense of T CD4+ lymphocytes can be an important confounding factor, since most of the treating physicians associate this finding with HIV/AIDS, as in this case. Importantly, up to 25% of patients with CVID present with low LT CD4+, without commitment of cellular immunity.

The goal of treatment is replacement of the humoral response by administering human immunoglobulin in order to reduce infectious and autoimmune complications, and the emergence of granulomas or malignancy. In Colombia different forms of human immunoglobulin are available, and they are authorized by INVIMA for use in patients with antibody deficiencies. The IVIG must be administered at a dose between 400-800 mg/kg every 3-4 weeks, depending on the valley levels and the clinical response; we also have subcutaneous human immunoglobulin (SubQ) for weekly application between 100- 200 mg / kg dose. The use of antibiotic prophylaxis is not clearly established; however, it is recommended the use of macrolides and quinolones; and for cases that present with low CD4+ T lymphocytes counts, it is recommended prophylaxis with trimethoprim/sulfamethoxazole.

Primary immunodeficiencies in adults must be a diagnostic possibility, and in Colombia, the technologies and methodologies to confirm the diagnosis are available. This case report aims to draw attention to a disease that requires a high index of suspicion, and the low reported prevalence is possibly due to underreporting and under diagnosis, which has an impact on the prognosis of the disease.
Referencias


