URTICARIA TREATMENT

596 Application of Intravenous Immunoglobulin for Treatment of Chronic Autoimmune Urticaria

Vladimir Alyoshkin, MD. Immunology, Moscow, Russia.

Background: To develop a patogenetically justified method for treatment of the autoimmune form of chronic urticaria.

Methods: 14 patients with autoimmune urticaria (18–60 y.o., time since disease onset ranging from 6 weeks to 5 years), having positive autoserum test results (7 mm or more in 11 patients, 4.5–6.5 mm in 3 patients) were treated with intravenous immunoglobulin for 4 days (50 mL of 5% immunoglobulin solution per day). Intravenous immunoglobulin of a fourth generation, containing 99.1 to 99.2% monomeric IgG, was applied.

Results: The treatment resulted in the rashes regressing in 13 (93%) patients after 3 to 4 days of treatment. All of the patients showed reduced autoserum test results (one of them was absolutely negative) 6 months after the end of treatment. Our investigations have shown a complete absence of clinical manifestations of urticaria within 1 year.

Conclusions: Intravenous immunoglobulin, containing only monomeric IgG, has proven highly efficient in treatment of chronic autoimmune urticaria. The above-mentioned reduced autoserum test results suggest fourth-generation intravenous immunoglobulin’s contribution to the pathogenesis of the disease.

X-LINKED AND COMMON VARIABLE IMMUNODEFICIENCY

597 Long Term Follow-up of Patients with Common Variable Immunodeficiency (Cvid) in Rio De Janeiro, Brazil: Clinical Phenotypes and Prognosis

Amanda Seba, MD, Norma de Paula Motta Rubin, PhD, Albertina Varandas Capelo, MSc, Eliane Miranda da Silva, MD, Mariza Campos de Magalhães, MSc, Fernando Samuel Sion, PhD, and Carlos Alberto Morais de Sa, PhD.

Clinical Immunology, Federal University of Rio de Janeiro State, Rio de Janeiro, Brazil; 2 Hematology, Federal University of Rio de Janeiro State, Rio de Janeiro, Brazil; 3 Internal Medicine, Federal University of Rio de Janeiro State, Rio de Janeiro, Brazil.

Background: CVID comprises a variety of clinical phenotypes that may influence the prognosis of the disease. Our goal was to investigate the clinical phenotypes and prognosis of a series of patients with CVID.

Methods: We evaluated 11 patients with CVID, according to the PAGID criteria in long-term clinical follow-up (> 10 years). Most patients were on regular use of intravenous immunoglobulin (IVIg), provided free of charge by the government. Clinical evaluation was performed monthly and exams every 6 months to 1 year, including immunological evaluation, hematologic, biochemical, autoimmune, stool, urine analysis, chest CT, abdominal ultrasound and specific investigations of infectious diseases and malignancies, when needed.

Results: The average follow-up was 21.9 years (12–34). Among the 11 patients, the mean current age was 39.8 years (16 to 62), 73% were female and 27% male. The age at symptoms onset ranged from 4 to 31 years (mean = 18) and diagnosis occurred between ages 11 and 47 (mean = 28). Most patients (55%) had the phenotype of infectious complications only, 27% had infections and immune thrombocytopenic purpura and 18% had infections and solid neoplasias. The most common infections were recurrent sinuses (100%), pneumonia (82%), giardiasis (36%) and tuberculosis (18%). None of the patients developed lymphoproliferative and / or inflammatory complications. With regard to immunological changes, we observed that 4 patients (36%) experienced an increase in CD8 T lymphocytes and inversion of CD4/CD8 ratio. Adherence to the use of IVIg was good in 50% of patients, fair in 38% and unsatisfactory in 12%. All patients have good quality of life, performing their routine activities of study, work and leisure.

Conclusions: In the population studied, the most frequent phenotypes were infectious complications or infectious complications + autoimmunity. Tubercolusis can be an important infectious complication in patients with CVID in endemic areas. The delay in the diagnosis of CVID, around 10 years, indicates the need to improve the diagnosis of PID in our country. With proper clinical management and good adherence to the use of IVIg, patients with CVID in developing countries may have survival and quality of life similar to those described in developed countries.

598 Quality of Life in Patients with Common Variable Immunodeficiency in the Department of Allergy and Clinical Immunology—Centro Medico Nacional Siglo XXI

Freya Helena Campos Romero, MD; Nora Hilda Segura Mendez, MD; Laura Mendoza, MD; and Nelva Guillén, MD.

Allergy and Clinical Immunology, Specialty Hospital, Centro Medico Nacional Siglo XXI, Mexico City, Mexico; and Allergy and Clinical Immunology, Mexico City, Mexico; 2 Allergy and Clinical Immunology, Specialty Hospital Centro Medico Nacional Siglo XXI, Mexico City, Mexico; 3 Allergy and Immunology, Mexico Distrito Federal, Mexico, and Allergy and Immunology, Specialty Hospital, Centro Médico Nacional Siglo XXI, México Distrito Federal, Mexico.

Background: Common variable Immunodeficiency (CVIDs) is characterized by a deficiency in antibody production and also generates enormous morbidity of school absenteeism and dropout or cause layoffs. All chronic diseases affect quality of life of patients, in this particular case CVIDs. The use of instruments like the SF - 36 provides information about the patients’ perception of their disease and its environment. The objective is to determine the quality of life of adult patients with CVID who receive replacement therapy through the use of SF-36.

Methods: We conducted a cross-sectional cohort study, which included all patients diagnosed with CVID in our hospital with approval from local research committee F-2011-3601-21. We analyzed the results with descriptive statistics and the SF-36 was used the method of Rand Group.

Results: Involving 11 patients with CVID, 4 men and 7 women, average age 30 years (18–53) years. The results show a reduction in quality of life of 65%, without gender difference. However in general we see that men are more affected the physical role 31% and women in the general health 40%. In mental health assessment, we found that in both genders is more affected in Vitality 55%, and Role Emotional least 84.72%. In relation to physical health, both genders showed greater involvement in aspects of general health 45% and less area affected is physical function 68 %.

Conclusions: CVID patients show a significant deterioration in their quality of life. It’s important to highlight that there are not statistically different variations in quality of life. But the general perception of gender does vary without establishing differences between mental and physical health.

599 Prevalence of Common Variable Immunodeficiency in Adult Patients Specialty Hospital—Centro Medico Nacional Siglo XXI - Ms—Mexico City

Nelva Guillén, MD, Nora Hilda Segura Mendez, MD, Laura Mendoza, MD, and Freya Helena Campos Romero, MD. Allergy and Clinical Immunology, Specialty Hospital, Centro Medico Nacional Siglo XXI, México City, Mexico.

Background: Common Variable Immunodeficiency (CVID) is the most common primary immunodeficiency (IDP), occurs primarily in adults between the second to fourth decades of life, without gender predominance. It is characterized by concentrations of immunoglobulins (Igs) of at least 2

S206 © 2012 World Allergy Organization

Copyright © 2012 World Allergy Organization. Unauthorized reproduction of this article is prohibited.
standard deviation (DS) of normal, recurring infections, some patients have autoimmune diseases and tumors, absence of isohemagglutinins and lack of production of antibodies (Ab) after vaccination and have excluded other causes of hypogammaglobulinemia.

Objective: Determine the prevalence of CVID in the Specialty Hospital—Centro Medico Nacional Siglo XXI, IMSS, Mexico City.

Methods: We included all patients with suspected diagnosis of CVID, clinical history, laboratory tests, determination of serum IgG and isohemagglutinins and were given a dose of gamma globulin 500 mg/kg/dose every 21 days. The results were analyzed with descriptive statistics. The protocol was approved by the local research committee.

Results: A total of 15 patients, 11 women and 4 men, mean age 34 years (± 11), 14 patients met all international standards and just only one patient has positive isohemagglutinins. The prevalence of CVID calculated according to the total population is that entitled in this hospital, 1, 520, 900, 1: 101, 393. In relation to the number of patients served by our service, the prevalence of 0.053% with a ratio of 1:2.533 right holders.

Conclusions: Our results show differences in relation to the published literature, prevalence in female sex ratio of 3:1, participating patients were diagnosed at the 34 years of average age, just only 2 patients were diagnosed before 20 years of age, unlike some of the international and national reports, we establish the definitive diagnosis according to international standards in 93% of cases evaluated. The prevalence of CVID obtained in our study is 1:101.393, higher than reported in international literature is 1:30.000 to 1:50.000, but similar to that reported in Spain in 1997 and Iran in 2006.

600 Infections in 14 Patients with Common Variable Immunodeficiency, Retrospective Study

Elizabeth Mendiesta,1 Leonel Gerardo Del Rivero, MD,2 and Nadia Aguilar3.
1 Allergy & Clinical Immunology, Specialty Hospital Medical Center XXI Century, Mexico City, Mexico; 2 Allergy and Immunology, Mexico City, Mexico; 3 Allergy & Clinical Immunology, Specialty Hospital Medical Center XXI Century, Mexico City, Mexico.

Background: Common variable immunodeficiency is a heterogeneous syndrome of primary antibody production failure. It affects 1 in 10000 to 50000 individuals, and is the most frequent primary immunodeficiency producing relevant clinical symptoms in adults and children. The hallmark of this disease is recurrent bacterial infections, usually of the respiratory and gastrointestinal tract. Onset is mainly in children aged 1 to 5 years, adolescents aged 16 to 20 years, and adults (fifth decade).

Methods: We assessed retrospectively recurrent infections in 14 patients with definitive diagnosis of CVID, for a period of 2 months through the review of their medical records.

Results: Ten patients were female (71.4%) and 4 were male (28.5%). The average age was 34 years. The average age of diagnosis of CVID was 27.5 years with an age range from 6 to 60 years. In 9 patients (64%) of the total studied CVID diagnosis was made in adulthood. All patients had a history of respiratory infection process in the following distribution: in 9 patients (64%) found a history of bronchiectasis, in 8 patients (57%) was found rhinosinusitis, and pneumonia; in 5 patients (35%) recurrent or chronic otitis media and one patient was a history of pulmonary tuberculosis. The lower urinary tract infection was found in 11 patients (78%), chronic diarrhea in 5 patients (35%), osteomyelitis in 1 patient.

Conclusions: Recurrent infections of the respiratory tract specifically low and high / or gastrointestinal infections should lead to systematic evaluation in which the primary immunodeficiencies are included as CVID.

REFERENCE

1. García JM, Gamboa P, De la Calle A, Hernández MD, Caballero MT. Diagnosis and Management of Immunodeficiencies in Adults by Allergologists J Investig Allergol Clin Immunol. 2010;20:185–194.

601 Autoimmune Thrombocytopenic Purpura Associated with Common Variable Immunodeficiency

Elizabeth Mendiesta,1 Leonel Gerardo Del Rivero, MD,2 1 Doctors in training, Mexico City, Mexico; 2 Allergy and Immunology, Mexico City, Mexico.

Background: Common variable immunodeficiency (CVID) is a condition characterized by antibody deficiency, and therefore susceptible to recurrent pyogenic infections, cancer and autoimmune diseases. It is a heterogeneous syndrome in primary immunodeficiencies and clinically the most important is often diagnosed in adulthood. Autoimmunity occurs in 5% of the general population, in patients with CVID the percentage increased to 20 to 48%, cytopenias being the most common cause of autoimmune in these patients. Autoimmune thrombocytopenic purpura and autoimmune hemolytic anemia are the most common autoimmune consequences, occurring in 5% to 8% of all patients with CVID. Some patients develop these disorders before the diagnosis of CVID.

Methods: We present the case of a woman of 45 year old, with a history of lower respiratory tract and urinary tract infections in recurrent Pulmonary Tuberculosis. Enter the program short-course treatment strictly supervised for pulmonary tuberculosis with appropriate response. Autoimmune thrombocytopenic purpura refractory to steroids (WWTP) for performing splenectomy.

Results: Anti DNA antibodies, anti nuclear, anti-protease, C. ANCA/PR3 antiimioperoxidasa, serology for hepatitis B, C, HIV negative. Serum immunoglobulins were as follow: IgG, 158 mg/dL (normal 700 to 1600), IgM, 55 mg/dL (normal 40–230), IgA, 36 mg/dL (normal 70–400), and, IgE, 38.7 IU/mL (normal 0–100) in more than 2 occasions with values below 2 standard deviations. CD4 T lymphocytes (19%) CD4/CD8 ratio (0.54).

Conclusions: Meets diagnostic criteria for Common Variable Immunodeficiency (CVID) and starting treatment with intravenous immunoglobulin at a dose of 400 mg/kg (every 21 days) with significant clinical improvement and has even managed to integrate into your daily activities. Today, he continues with danazol for WWTP. Therefore, CVID is necessary to consider in the differential diagnosis of autoimmune thrombocytopenic purpura and autoimmune hemolytic anemia in adults (1).

REFERENCE

1. Miguel Park A, et al. Lancet. 2008;372:489–502.

602 Cvid: A Common but Still Underdiagnosed Disease

Claudia Gallego, MD,1 Sandra González-Díaz, MD, PhD,1 Maria del Carmen Zarate, MD, PhD,1 Alfredo Arias-Cruz, MD,1 Diego García-Calderin, MD,1 Karla Yanneth Mejía Salas, MD,2 Maricruz Calva, MD,3 and Luis Alfredo Dominguez Sansores, MD3.
1 Regional Centre of Allergy and Clinical Immunology, University Hospital, Monterrey, Mexico; 2 Allergy and Clinical Immunology, Regional Centre of Allergy and Clinical Immunology, Monterrey, Mexico; 3 Regional Centre of Allergy and Clinical Immunology, Monterrey, Mexico.

Background: Among the more than 150 different forms of Primary Immunodeficiency Diseases (PID) the CVID is the most common symptom-atic primary immunodeficiency, present mainly in adults. There is a failure of B cells to develop and differentiate into plasma cells; at consequent a reduction of the production of one or more isotypes of antibody can also affected Cell-mediated immunity. Common manifestations included recurrent bacterial infections, that typically involve the upper and lower respiratory tract. Some patients are highly prone to autoimmune manifestations, lymphoid hyperplasia, and tumors.

Methods: We presented 3 cases of CVID with a variety of clinical presentation, evolution and complications related to delayed diagnosis.