

Acute Rejection of a Kidney Transplant in a Patient With Common Variable Immunodeficiency: A Case Report

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ABSTRACT

Common variable immunodeficiency is a primary immunodeficiency characterized by hypogammaglobulinemia and recurrent bacterial infections. We report a case of a 44-year-old male patient with end-stage renal disease and an established diagnosis of common variable immunodeficiency who underwent a living unrelated kidney transplant. He remained nearly infection free on maintenance immunoglobulin replacement. However, his posttransplant course was complicated by acute rejection that ultimately led to allograft loss. This case illustrates the challenge of transplantation in this patient population because of the delicate balance that must be achieved between maintaining adequate immunosuppression and minimizing the risk of infection.

COMMON VARIABLE IMMUNODEFICIENCY (CVID) is a primary immunodeficiency that is defined by markedly reduced levels of IgG, low levels of IgM and IgA, poor or absent response to immunizations, and the absence of any other defined immunodeficiency state [1]. CVID is characterized by impaired B-cell differentiation that leads to defective immunoglobulin production and severe antibody deficiency. In the majority of cases, the mechanism for the defect remains poorly understood. In addition, evidence of immune dysregulation and T-cell dysfunction is common [2,3]. This disease is relatively common, with the incidence estimated at 1 per 75,000 live births. The disease is usually diagnosed during the third or fourth decades of life [3,4]. The primary clinical manifestations of CVID are recurrent bacterial sinopulmonary infections caused by encapsulated bacteria. Other clinical manifestations of CVID are heterogeneous, ranging from recurrent viral infections to autoimmune disorders to neoplasms [2,5]. Renal disease is not considered to be a part of the disease spectrum. The mainstay of treatment for CVID is regular replacement of IgG to reduce the incidence of infections. IgG can be administered via the intravenous or subcutaneous route. A target IgG trough level of 600 to 900 mg/dL typically minimizes the incidence of any further recurrent bacterial infections [6].

There are a limited number of reports in the literature regarding patients with CVID who have undergone solid organ transplantation (Table 1). Due to the risk of infection and malignancy (particularly lymphoma) with further immunosuppression, there is hesitancy in pursuing transplantation in these patients. This hesitancy may be more pronounced in patients with end-stage renal disease due to the availability of dialysis as a treatment alternative. To our knowledge, only 1 other report of kidney transplantation occurring in a patient with CVID has been published [14]. In that particular case, CVID was diagnosed retrospectively after the patient presented with numerous infections posttransplant.

In the present article, we describe the clinical course of a patient with an established diagnosis of CVID who underwent living unrelated kidney transplantation.

CASE REPORT

A 44-year-old white male patient with end-stage renal disease due to an unknown cause was diagnosed in November 2010 with CVID

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Table 1. Reported Cases of Solid Organ Transplantation in Patients With CVID

Author	Reason for Organ Failure	Timing of CVID Diagnosis	Organ Transplanted	Induction Immunosuppression	Maintenance Immunosuppression	Rejection Episodes	Complications
Smith et al [7]	HCV	Before transplant	Liver	Not reported	Cyclosporine, azathioprine, steroids	No	Recurrent HCV, sepsis, ARDS
Smith et al [7]	Hepatic granuloma	Before transplant	Liver	ATG	Azathioprine, steroids	Acute cellular rejection on day 6 posttransplant	Sepsis, ARDS, thrombocytopenia
Montalti et al [8]	HCV	Before transplant	Liver	Basiliximab	Tacrolimus, everolimus, steroids	No	None
Montalti et al [8]	HBV	Before transplant	Liver	Not reported	Tacrolimus, MMF, steroids	Acute rejection 6 months posttransplant	CMV
Chen and Cameron [9]	Autoimmune hepatitis	Before transplant	Liver	Basiliximab	Tacrolimus, MMF, steroids	No	Cerebral aspergillosis
Burton et al [10]	Bronchiectasis	Before transplant	Lung	ATG	Cyclosporine, azathioprine, steroids	Acute cellular rejection at weeks 4 and 6 posttransplant	CMV, sepsis, pulmonary aspergillosis and other organisms
Gow and Mutimer [11]	HCV	Before transplant	Liver	Not reported	Cyclosporine, azathioprine, steroids	Mild acute cellular rejection on day 8 posttransplant	Severe “fibrosing” cholestatic hepatitis
Murakawa et al [12]	Fulminant hepatitis	Before transplant	Liver	Not reported	Tacrolimus	Acute cellular rejection 5 years posttransplant	Not reported
Murakawa et al [12]	Hepatitis	Before transplant	Liver	Not reported	Tacrolimus, steroids	Five episodes of severe acute cellular rejection	Recurrent hepatitis
Hill et al [13]	Bronchiectasis and emphysema	Before transplant	Heart/Lung	Not reported	Cyclosporine, azathioprine, steroids	Minor episode of rejection at 2 months posttransplant	Not reported
Hogan et al [14]	Unknown	After transplant	Kidney	ATG	Cyclosporine, azathioprine, steroids	Four acute cellular rejection episodes in the first year posttransplant	Oral candidiasis

Abbreviations: ARDS, acute respiratory distress syndrome; ATG, antithymocyte globulin; CMV, cytomegalovirus; CVID, common variable immunodeficiency; HBV, hepatitis B virus; HCV, hepatitis C virus; MMF, mycophenolate mofetil.

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