# Successful Treatment with Rituximab in a Patient with Castleman's Disease Complicated by Systemic Lupus Erythematosus and Severe Autoimmune Thrombocytopenia

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To the Editor: A 44-year-old man with no remarkable past medical history, was admitted with 1-month history of fever and lymphadenopathy. He also had night sweats, malaise, arthralgia, urine volume reduction, abdominal distention, and weight loss. Physical examination revealed swollen lymph nodes in the neck, axillary fossae and inguinal grooves, palpable liver and spleen, shifting dullness and edema. Laboratory investigation showed anemia (hemoglobin 89 g/L) and severe thrombocytopenia (platelet $9 \times 10^9$/L), positive urine red blood cell and urine protein (0.68 g/24 h), elevated creatinine (180 μmol/L), elevated erythrocyte sedimentation rate (ESR) (35 mm/h), elevated gammaglobulins (IgG 19.3 g/L), and low albumin level (27 g/L). Antinuclear antibody (ANA, speckled pattern, 1:160), anti-dsDNA, and anti-SSA antibodies were positive. Complement levels were reduced. Echocardiogram showed normal cardiac function with a mild pericardial effusion. A computed tomography scan revealed bilateral pleural effusions, enlarged lymph nodes in the mediastinum and the retroperitoneal region, hepatosplenomegaly, and ascites. Paracentesis yielded yellow, clear fluid with a serum-to-ascites albumin gradient of 6 g/L. Cultures were negative. Bone marrow aspiration showed increased large and immature megakaryocytes. Lymph node biopsy suggested Castleman's disease (CD) of the hyaline vascular variant. A diagnosis of multicentric CD complicated by systemic lupus erythematosus (SLE) and severe thrombocytopenia was confirmed. Patient was treated with two courses of CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) regimens as well as intravenous immunoglobulin (Ig) infusion, steroids, danazol, and thrombopoietin. However, he still had a temperature, low platelet count ([5–7] $\times 10^9$/L) despite a partial shrank of lymph nodes, liver and spleen. Rituximab (R) were then given (600 mg/week for four doses) and the patient improved significantly after the treatment [Figure 1]: Temperature returned normal; peripheral lymph nodes as well as liver and spleen became unpalpable; hemoglobin up to 125 g/L, platelet $110 \times 10^9$/L; urine blood (−); urine protein (−) (0.18 g/24 h); ESR 20 mm/h; creatinine 93 μmol/L; albumin 36 g/L; IgG 5.58 g/L; ANA, anti-dsDNA, anti-SSA antibodies were negative. Imaging studies revealed disappearance of pleural effusion and ascites, absence of enlarged lymph nodes and normalized liver and spleen. Three courses of R-COP were given as maintenance therapy. After that, the patient went home without need for further steroids or immunosuppressants therapy. He remained disease-free and maintained good functional status on his last follow-up at 7 years after initial diagnosis.

CD is a heterogenous group of lymphoproliferative disorders characterized by a hyperplastic-reactive process involving the immune system. CD complicated with SLE is a rare clinical condition with only a few cases[1-3] reported. Although autoimmune thrombocytopenia (AITP) was seen in nearly 50% of these patients, the severity was mostly mild to moderate, while severe thrombocytopenia at the onset of disease (before chemotherapy) has not been previously reported. Thrombocytopenia was our major concerns during treatment as it did not respond to potent chemotherapy. Although the use of rituximab in SLE is still controversial, its efficacy in CD[4] and AITP[5] has been reported. Moreover, its success in treating our patient suggests a potential role of rituximab in treating similar patients.

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## Conflicts of interest

There are no conflicts of interest.

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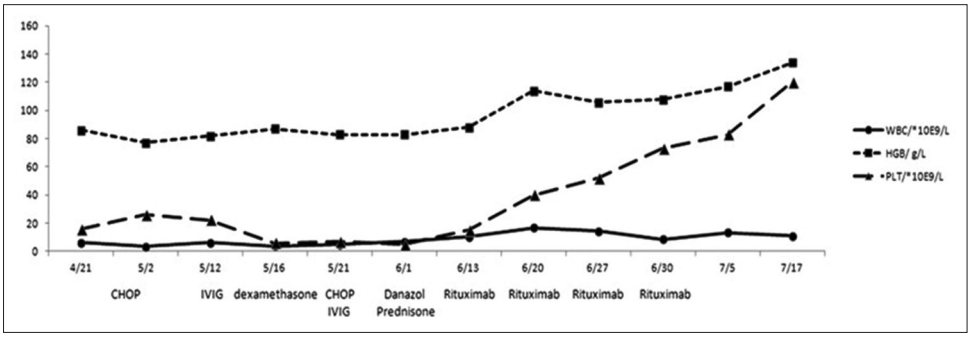


**Figure 1:** Treatment response of blood routine test in a patient with Castleman's disease complicated with systemic lupus erythematosus and severe autoimmune thrombocytopenia.

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