AB003. OS01.03. New proposed treatment for pure red cell aplasia thymoma-related

Giovannella Palmieri[1], Margaret Ottaviano[1], Stefano De Falco[2], Giuliano Palumbo[1], Carmen Forino[1], Paola Fiorillo[3], Elide Matano[1], Vincenzo Damiano[1]

[1]Department of Clinical Medicine and Surgery, [2]Oncology Unit, Department of Clinical Medicine and Surgery, University of Naples Federico Ii, Naples, Italy; [3]Department of General and Specialistic Surgery, University of Campania Luigi Vanvitelli, Naples, Italy

Background: Patients with thymic epithelial tumors (TETs), compared with controls, are more likely to have an autoimmune disease during their lives (32.7% *vs.* 2.4%; P<0.001). The association of several parathymic entities, including Good syndrome (GS) and haematological autoimmune disease is not uncommon. Pure red cell aplasia (PRCA) aplastic, haemolytic, and pernicious anemia are reported in 50% to 86% of patients. Here we describe the clinical features and the treatment outcomes of patients with GS and PRCA, proposing a new treatment strategy that combines the prolonged control of both autoimmunity and immunodeficiency.

Methods: We retrospectively analysed data of patients with thymoma and both diagnosis of GS and PRCA, who were admitted to our Institution over a 21-year period from October 1995 to September 2016. GS was diagnosed through the evidence of hypogammaglobulinemia and a marked decrease of CD19 and CD20 positive B lymphocytes in the peripheral blood, while the PRCA diagnosis was made by the suppression of the erythroid series in the bone marrow (<5%). Treatment outcomes were registered.

Results: With a median age of 44 years (range, 33–65 years) and a ratio male/female of 1/1, 14 patients were included in the study, of whom 3 received neo-adjuvant chemotherapy before surgery, 6 underwent radical thymectomy and the last 5 were candidate to first line therapy for unresectable disease. The histotype B2 was the most diagnosed (57%). Before starting any immunosuppressive treatment, all the patients received therapy for GS with administration of intravenous immunoglobulin (IVIG) at 500 mg/kg per day for 5 days/monthly and prophylactic antibiotic and anti-mycotic treatment. The best outcome with the faster and prolonged PRCA resolution has been recorded in 9 patients treated with a combination therapy of prednisone at the dosage of 0.6–1 mg/kg/daily, long acting octreotide at the dosage of 30 mg every 4 weeks and oral cyclosporine at a daily dose of 200 mg. Five patients, among those treated with the combination, are still alive and without PRCA recurrence, the others four died for progressive disease. The last five patients, managed with more aggressive immunosuppressive agents, such as tacrolimus and alemtuzumab, despite the GS therapy, died due to fatal infections.

Conclusions: Taking into account our experience, we recommend the use of a combination therapy for GS and PRCA control. More aggressive immunosuppressive therapy, such as alemtuzumab and tacrolimus, could be avoided due to the risk of fatal infections. High expertise to manage these patients is strongly required.

Keywords: High expertise; pure red cell aplasia (PRCA); combined treatment; good syndrome

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