

ANCA-negative eosinophilic granulomatosis with polyangiitis in a young adult treated with Reslizumab

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Eosinophilic granulomatosis with polyangiitis (EGPA), previously known as Churg–Strauss syndrome, remains a rare but complex disease characterized by asthma and eosinophilic tissue infiltration of multiple organ systems. Currently, anti-IL-5 therapies represent safety and efficacy in the treatment of EGPA. Herein, we report the case of a young patient with EGPA for implementation of reslizumab (Cinqair®; Teva, North Wales, PA, USA) due to the difficulty in tapering treatment with systemic corticosteroids. A 24-years-old female was referred to the Emergency Department with dyspnea and purpura in both lower limbs for one month. She had been diagnosed with asthma and chronic rhinosinusitis with nasal polyp 1 years earlier. Blood tests showed an eosinophilic count of 11,379 cells/ μ L with negative Antineutrophil cytoplasmic antibodies (ANCA). Chest CT scan showed bilateral pulmonary peripheral opacities. Skin biopsy performed from purpura showed vasculitis with eosinophilic infiltration. These clinicopathological findings led to the diagnosis of EGPA. She was initially started on methylprednisone 1 mg/kg per day and obtained acceptable asthma control as well as remission of purpura and peripheral eosinophilia. At the 6-month follow-up, with daily 4mg oral methylprednisone, clinical manifestations were in remission and eosinophils were in the normal range. However, tapering of corticosteroids could not be further accomplished without rebound of the circulating eosinophil count up to 10,154 cells/ μ L and new asthma exacerbation. Because of the side-effect burden and ineffective improvement of oral corticosteroids, a fixed dose of 100 mg reslizumab was intravenously administered at monthly intervals. Three months after starting treatment with reslizumab, her blood eosinophils showed below 200 cells/ μ L and daily oral corticosteroids had been completely suspended. Moreover, her respiratory symptoms, lung function parameters and fractional exhaled nitric oxide levels were improved, and she has reported no further exacerbations and can be successfully stepped down controller treatment. Reslizumab is still ongoing as the maintenance therapy, 100 mg monthly intravenously.

