A case of Churg-Strauss syndrome with hemorrhagic cystitis after prolonged oral cyclophosphamide therapy

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Churg-Strauss syndrome (CSS) is a necrotizing vasculitis involving small to medium-sized vessel characterized by asthma, lung infiltrates, extravascular necrotizing granulomas, and hypereosinophilia. The use of cyclophosphamide (CYC) has been advocated in high risk patients or patients with neuropathy to prevent or reduce mortality and disability. When necessary, intravenous pulse CYC is recommended to reduce side effect, but sometimes daily oral CYC treatment is required for pulse treatment resistant cases. Hemorrhagic cystitis associated with daily dose of oral CYC and it has rarely been reported in the past 20 years. Herein, we report a 36-year-old man with hemorrhagic cystitis who received 25 mg of oral CYC daily to treat CSS with severe peripheral neuropathy (total CYC dose was 137g for 6 years). Patient initially presented with asthma, paranasal sinusitis, lung infiltrate on chest computed tomography, and peripheral eosinophilia, and a biopsy demonstrated the accumulation of eosinophil in the perivascular and peribronchial septum in the lungs and leukocytoclastic vasculitis with eosinophil infiltration on the involved skin. Initially high dose of systemic glucocorticoid was administered intravenously and tapered to oral prednisolone. Eighteen months later, patient develop left foot drop. Two cycles of intravenous CYC pulses were administered with steroid pulse therapy but patient complained of newly developed both wrist and right foot drop with paresthesia. The neurologic manifestations were improved after oral CYC treatment (1.5 mg/kg/day was provided initially and maintained at 0.25 mg/kg/day for over 6 years). Six years after oral CYC treatment, hemorrhagic cystitis developed suddenly. No other side effect occurred for 6 years. After the discontinuation of oral CYC, hematuria disappeared. The maintenance of oral CYC was effective for CYC pulse treatment-refractory neuropathy. Although it is known that duration and daily dose of CYC are important factors in development of hemorrhagic cystitis. But hemorrhagic cystitis might occur at low daily dose of CYC. Routine urinalysis and history-taking are crucial for patients receiving oral CYC for a long period even in the absence of other side effect.