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SCORTEN on the third day of hospitalization can be a predictive factor for Korean Stevens-Johnson syndrome and toxic epidermal necrolysis patients

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Backgound: Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are very rare but potentially life-threatening conditions with grave prognosis. SCORTEN has been developed to predict the risk of mortality in SJS/TEN patients. However, the clinical usefulness of SCORTEN is still in controversy. Objective: To evaluate the clinical usefulness of SCORTEN in Korean population. Methods: Medical records of 48 patients with SJS or TEN who visited Seoul National Hospital University from Jan 1, 2005 to Dec 31, 2010 were reviewed. Vital signs and laboratory findings on the day of admission, 3rd hospital day, and 5th hospital day were collected and the clinical course of the patients including mortality, hospital stay, ICU admission, and the presence of sequelae were reviewed. SCORTEN score of hospital day 1 (SC1), 3 (SC3) and 5 (SC5) were calculated and SCORTEN score using maximal available values during first 5 days of admission (SCmax) was also evaluated. Results Of 48 patients enrolled male to female ratio was 1:1. Median age was 47 (0-86) years old and there were 37 (77.1%) SJS patients and 11 (22.9%) TEN. Four (8.3%) of 48 died during their hospital courses. As not all the subjects had their laboratory test results for the days investigated, patients who had their test results on that day were only included in the analysis. SC1 was calculated in 44 patients, SC3 in 18, SC5 in 16, and SCmax in 48, respectively. Among the scores, SC3 showed significant association with the mortality (p for trend=0.022), but not with other clinical outcomes such as sequelae, ICU admission or duration of hospital stay. SCmax showed borderline significance for mortality (p for trend=0.073) and ICU admission (p for trend=0.062). SC1 and SC5 did not showed significant association with none of clinical outcomes. Conclusion: This is the first study to evaluate usefulness of SCORTEN in Korean SJS or TEN. SCORTEN score calculated on the third day of hospitalization could be a predictive factor of mortality for Korean SJS/TEN patients. This research was supported by a grant (09182KFDA889) from Korea Food &Drug Administration in 2011.

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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.