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Case Report

A case of Stevens–Johnson syndrome with gross hematuria☆

Dear Editor,

Stevens-Johnson syndrome (SJS) is potentially fatal disorder, confirmed on the basis of most widely accepted Bastuji-Garin definition, which included widespread macules or flat atypical targets, and detachment of skin involving at least two mucous membranes (Bastuji-Garin et al., 1993). Involvement of the buccal, genital and ocular mucosa occurs in most cases (Harr & French, 2012). In some cases the respiratory and gastrointestinal tracts are also affected, and of corneal and colonic perforation (Baccaro et al., 2014; Lebargy et al., 1997; Revuz et al., 1987). Baccaro LM and his colleagues had reported a case of toxic epidermal necrolysis(TEN) with acute renal failure secondary to ureteral obstruction and perforation (Baccaro et al., 2014). Herein, we experienced a case of SJS with ureteropelvic mucosa involved, resulting in gross hematuresis.

A 27-year-old Chinese woman with a healthy medical history admitted to our hospital. Physical examination revealed a generalized purpuric macules with flaccid blisters filled with serous liquid was observed on the face and trunk of the patient, and extremities with a few purpuric macules. Nikolsky's sign was confirmed, and epidermal detachment below 10% of the body surface area(BSA). Erosive lesions were found in her ocular, oral, pharyngeal, and genital mucosae. Erosive lesions prevented the patient from opening her eyes. The patient also had difficulty speaking and eating. The patient went on to develop swollen and hemorrhagic lips and eyelids that crusted with black eschars (Fig. 1). What's more, the color of urine was dark red.

Initial laboratory blood studies revealed no abnormality seen in routine blood test, hemoglobin was 120g/L (N=115–150g/L). Urinalysis showed a red blood cell (RBC) count of 40058.1/u. The patient's platelet count and coagulation profile were normal. Blood creatinine and urea nitrogen levels were within reference values. Liver function test results showed abnormal [AST, 368.9 U/L(N=13–35 U/L); ALT, 470.1 U/L(N=7–40U/L)). Histologic examination of the skin biopsy specimens found epidermis necrosis, and the direct immune fluorescence(DIF) staining was negative. The diagnosis of SJS was made. After two days, hemoglobin value was down to 86 g/L, and the bleeding received 500mL within two days.

Further examinations were performed to clarify the source of gross hematuria. Gynecological examination and laboratory tests ruled out disorders of obstetrics and gynecology. Examination of the urinary sediment with the phase-contrast microscope revealed less than 40% dysmorphic red cells, more than 60% monomorphic pattern exists, and erythrocyte casts were absent, which indicated the observed hematuria was a urological cause (Huussen et al., 2004). Plain and enhanced CT of whole abdomen was performed and showed thickening and enhancement of the walls of the patient's renal pelvis and ureter (Fig. 2), without any other abnormalities seen. Continuous bladder irrigation was performed to prevent blood clot formation.

Patient was administered high-dose intravenous immunoglobulin, methylprednisolone therapy, and supportive treatment. Her lesions of cutaneous were completely healed, And she was able to completely open her eyes and consume soft food, and liver function was improved, meanwhile the urine was deemed normal in color and urinalysis indicated normal without occult blood or RBC and left hospital. Until now, the patient was followed up for about two years, and she is in good health.

SJS and TEN are two entities of a similar immune modulated disorder affecting mucocutaneous membranes. differentiated principally by surface area and severity of the disease. Our case indicated that ureteropelvic mucosa could be damaged in the disease of SJS and resulted in gross hematuresis, even worse, acute renal failure because of extensive ureteral mucosal sloughing (Baccaro et al., 2014). The disease of SJS-TEN is a systemic disorder, which requires doctors to diagnose and treat it more carefully.

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☆ **Conflict of interest:** The authors declare having no conflicts of interest regarding the conduct or outcomes of this study.

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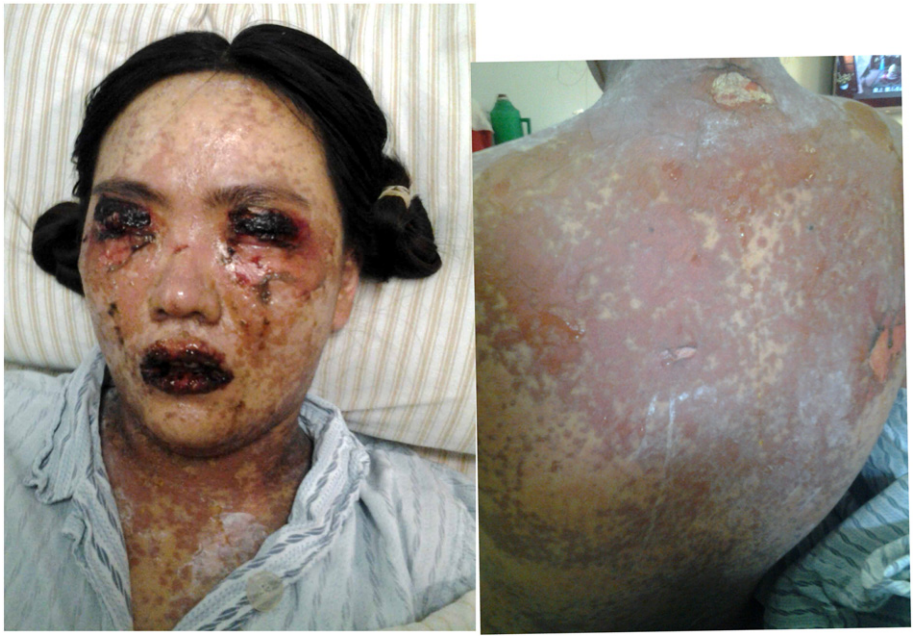

Fig. 1. The patient's lips and eyelids covered with black eschars. generalized purpuric macules and erosive lesions on the patient's face and neck and trunk.

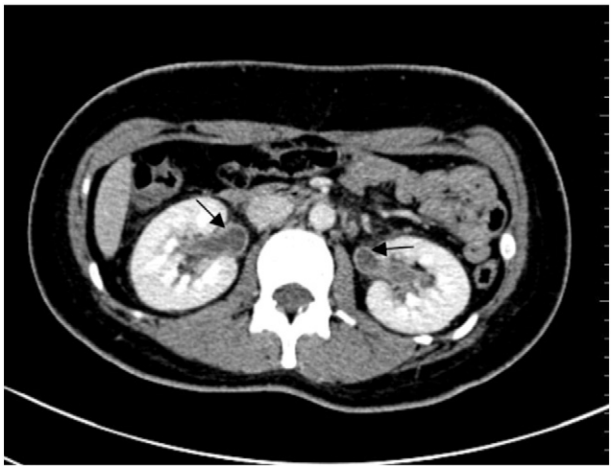

Fig. 2. Thickening and enhancement of the walls of the patient's renal pelvis and ureter.

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