Severe Large-Intestine Involvement in Adult-Onset Henoch-Schönlein Purpura: Successful Treatment with Factor XIII Concentrate

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Dear Sir,

Henoch-Schönlein purpura (HSP) is a systemic vasculitis of the small vessels of the skin, joints, gastrointestinal (GI) tract and kidney. GI involvement occurs in 50–75% of patients [1]. The duodenum is the predominant lesion, and HSP may also affect the esophagus, stomach, and colon. It is rare that the stricture occurs in the large intestine, as in our patient.

Fig. 1. Colonoscopy shows remarkable mucosal redness, edema, irregular ulceration, and a stricture in the descending colon.

A 24-year-old man presented with a palpable purpuric skin rash over the lower limbs on the day following onset, associated with bilateral ankle pain and swelling. He also had left lower abdominal pain. Five days later, the abdominal pain became intense concomitant with the appearance of bloody stools, and the patient was hospitalized. On examination, initial leukocyte count was $15,400/\text{mm}^3$, hemoglobin was 14.4 g/dl and C-reactive protein was 8.3 mg/dl. There was microscopic hematuria but no proteinuria. Skin biopsy showed evidence of leukocytoclastic vasculitis and IgA deposits were seen on immunofluorescence. Upper endoscopy revealed some mucosal redness in the gastric fornix. Colonoscopy revealed remarkable mucosal redness, edema, irregular ulceration, and a stricture in the descending colon (fig. 1). The obstructed segment prevented colonoscopic passage. The patient was diagnosed as having HSP based on the American College of Rheumatology criteria [2]. He was given intravenous corticosteroids (prednisolone, 1 mg/kg/day), but after 2 weeks of treatment, his symptoms did not resolve. Additional laboratory studies revealed his plasma level of factor

XIII was less than 40%, so we started treatment with factor XIII concentrate (20 ml a day for 3 consecutive days). With these measures, he recovered within 2 weeks. We succeeded in treating severe large-intestine involvement in a HSP patient by factor XIII concentrate.

References

- 1 Saulsbury FT: Clinical update: Henoch-Schönlein purpura. *Lancet* 2007;369:976–978.
- 2 Mills JA, Michel BA, Bloch DA, Calabrese LH, Hunder GG, Arend WP, Edworthy SM, Fauci AS, Leavitt RY, Lie JT: The American College of Rheumatology 1990 criteria for the classification of Henoch-Schönlein purpura. *Arthritis Rheum* 1990;33:1114–1121.