●〈症例〉

A case of Churg–Strauss syndrome diagnosed by characteristic skin lesions and gastrointestinal involvement

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〔Key Words〕 hypereosinophilia, gastrointestinal involvement

和文要旨

症例は71歳男性。主訴は下痢、腹痛、嘔気。既往歴に気管支喘息、心房細動。2004年5月下旬より膨隆し始めた紫斑性丘疹が下肢中心に多数みられ、血液検査にて好酸球増多を認めたため、精査加療目的で入院となった。入院直後より呼吸困難、3病日より下肢しびれ感が出現し歩行困難となった。上部消化管内視鏡では体部から幽門部にかけて地図状潰瘍が多発していた。下部消化管内視鏡では直腸からS状結腸および回腸終末部に粘膜の発赤と浮腫を認めた。いずれの生検組織も粘膜に優位に好酸球の浸潤があり、血管周囲の肉芽種性変化がみられた。皮膚生検組織は毛細血管周囲および血管壁へのリンパ球浸潤を認め血管炎の所見であった。以上よりCSSと診断しステロイド投与を行った。その結果、好酸球数は速やかに正常範囲内に低下し、消化器症状の改善も得られた。CSSは皮疹、神経症状で発症することが多いが、消化器病変の合併も少なくない。本症のように皮疹がみられ、消化器症状が続発する場合、本症も念頭に置き検査する必要があると思われる、若干の文献的考察を加えて報告する。

Table 1 Blood laboratory findings

WBC	18400 /uL	AST	52 U/L	BUN	8.5 mg/dL
Neut	9 %	ALT	41 U/L	Cre	0.78 mg/dL
Lymph	7 %	T-Bil	1 mg/dL	Na	139 mEq/L
Mono	0 %	LDH	298 U/L	K	3.9 mEq/L
Eosi	84 %	ALP	1188 U/L	Cl	102 mEq/L
Baso	0 %	γ-GTP	129 U/L	Ca	9.3 mg/dL
RBC	508 10/uL	CPK	93 U/L	IgE	832 IU/mL
Hb	15.3 g/dL	B-AMY	85 U/L	IL-5	350 Pg/mL
Ht	47 %	Tch	183 mg/dL	MPO-ANCA	11 EU
Plt	21.9 10/uL	PG	92 mg/dL	PR3-ANCA	10 EU
TP	7.4 g/dL	CRP	2.5 mg/dL

Fig. 1 Itchy purpuric papules on the lower limbs

Case Report

In June 2004, a 71-year-old man was admitted to the hospital complains of abdominal pain, watery diarrhea, and itchy purpuric papules all over the body (Fig. 1) for the past 3 days. On the evening of the admission day, he became aware of shortness of breath. He had been suffering from asthma and allergic rhinitis for several years. His EKG showed atrial fibrillation, which was diagnosed several years earlier. On physical examination, his temperature was 37.6°C; pulse rate, 76 beats/min; and blood pressure, 126/90 mmHg. He had numerous papular purpuric lesions almost all over the body. Auscultation of the chest revealed coarse crackles. Bowel frequency was more than ten times a day. On the third day in hospital, he complained of paresthesia and weakness in both lower extremities. Tendon reflexes were absent. Blood laboratory findings are listed in the (Table 1). The most pertinent findings were high-level leukocytosis 18,400/uL with 84% eosinophils; IgE, 832 IU/mL; and IL-5, 350 pg/mL. Chest X-ray showed infiltration of the right lower lobe. Parasites and other organisms were not detected in stool samples. Skin biopsy results showed inflammatory vasculitis.

Gastrointestinal endoscopy revealed multiple irregular gastric ulcers extending from the gastric body to the antrum (Fig. 2-a). Gastric submucosal biopsy showed granulomatous vasculitis with eosinophils (Fig. 3). On colonoscopy, the mucosa appeared red and edematous with erosive lesions extending from the rectum to the sigmoid colon and red patchy lesions located at the end of the ileum (Fig. 2-b). Rectal biopsy revealed severe eosinophilic infiltration of the mucosa and submucosa.

All these results confirmed the diagnosis of CSS. The patient received 60 mg/day prednisolone intravenously from the third day in hospital. After prednisolone administration, the eosinophil count decreased to the normal levels on the fifth day in hospital. With the exception of paresthesia in lower limbs,

Fig. 2-a Gastroscopic finding in the antrum; the irregular ulcers were surrounded by edematous gastric mucosa.

Fig. 2-b Colonoscopic finding in the cecum; the ileocecal valve was red and edematous.

all clinical complaints were gradually relieved. The prednisolone dose was tapered to 30 mg/day at discharge from the hospital.

To date, the patient has been followed-up as an out-patient. Currently he takes 5 mg/day of prednisolone. However the paresthesia on the lateral side of the right lower limb and flat purpuric lesions persist.

Discussion

CSS is an idiopathic multisystem vasculitis involving small and medium-sized arteries in patients with bronchial asthma or allergic sinusitis. The characteristic histologic features are granulomatous vasculitis, eosinophilic infiltration of tissues, and extravascular granuloma. Marked peripheral eosinophilia is the rule at the diagnosis of CSS. Clinical features of CSS depend on the organs involved, however most patients report nonspecific manifestations such as fever, malaise, and weight loss. The skin and peripheral nerves are most often affected, skin lesions include purpuric nodules, and peripheral nerves involvement results in paresthesia, paresis and paralysis¹⁾.

In this case, the patient chiefly showed gastrointestinal (GI) tract manifestations such as severe abdominal pain along with characteristic skin lesions and remarkable hypereosinophilia. CSS was suspected because of the presence of these incipient clinical features. However every incipient clinical finding is not always compatible with the diagnosis of CSS. We studied the early clinical manifestations of CSS in patients. We reviewed 120 patients reported in *Japana Centra Revuo Medicina* (Igaku-chuou-zasshi) from January 2000 to April 2005 (**Table 2**). The mean age of onset of CSS is 50.5 years with a male-to-female ratio of 1:1.26. The disease can occur at any age of with the possible exception of infants²⁾. Hypereosinophilia was observed in all cases. GI complaints were reported in 36 patients; 30% patients with CSS experi-

Fig. 3 Gastroscopic biopsy results showed granulomatous angitis with severe eosinophilic infiltration of submucosa

Table 2 Summary clinical findings of CSS: Literatures in *Japana Centra Revuo Medicina* (Igaku-chuou-zasshi) from January 2000 to April 2005

• Male-to-female ratio	1:1.26
• Mean age of onset	50.5 years (13–77 years)
• Clinical manifestations related to organ system involvement at the incipient stage	
Organ system	incidence percent
Gastrointestinal tract	36 30
Skin	25 21
Peripheral nervous system	65 54
Pulmonary	15 12
Cardiac	4 3
Gastrointestinal tract	incidence percent
Stomach	17 57
Duodenum	4 13
Small intestine	19 63
Colon	16 53

enced abdominal pain, nausea, diarrhea, and hematochezia. Other studies in literature reported values of 17%³⁾ and 50%⁴⁾. Based on these results, it is considered that GI tract involvement is not rare at the incipient stage in patients with CSS. However, only seven patients suffered from both skin lesions and GI tract involvement. The diagnosis of CSS should be considered in patients with GI tract involvement associated with hypereosinophilia.

Upper and lower endoscopic findings of CSS included characteristic multiple irregular ulcers, edematous mucosa, and patchy mucosal erythema^{5,6)}. Gastric lesions, particularly, ulcers, extend from the gastric body to the antrum; colonic lesions are typically located on the left-side. The findings of our case were compatible with these characteristic findings of CSS. Small intestinal lesions tend to be deep and result in perforations. In our study, small intestinal perforations were observed in 16 of the 19 patients with small intestinal manifestations. The mortality rate of CSS was 10.8%; 13 patients died due to CSS. The leading cause of death was GI tract hemorrhage and cardiac disease (5 patients each). The prognosis of treated CSS is therefore more likely to be related to the involvement of the GI tract and heart⁷⁾.

Involvement of the GI tract is not rare in CSS, and it is the leading cause of death. Therefore, it is important that CSS be suspected when GI tract manifestations are associated with hypereosinophilia.

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皮疹により診断しえたChurg-Strauss症候群の1例

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