Asymmetrical Polyneuropathy Associated with Churg-Strauss Syndrome

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Abstract

A 26-year-old man who presented with fever, leg edema, weakness and numbness of both hands and feet and painless palpable purpura was reported. He had had a history of asthma and arthalgia before this admission. Physical examination revealed distal asymmetrical polyneuropathy. Electrodiagnostic study was consistent with polyneuropathy. Skin biopsy revealed vasculitis with prominent eosinophilic infiltration. His motor power was markedly improved by prednisolone treatment.

Key word : Asymmetrical Polyneuropathy, Churg-Strauss Syndrome

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J Med Assoc Thai 2001; 84: 907-909**

Churg-Strauss syndrome (CSS) or allergic angiitis and granulomatosis is an uncommon disease. It is characterized by vasculitis of the multiple organ system, particularly the lungs. Of the neurological involvement, neuropathy is the most common manifestation which usually presents with mononeuritis multiplex[1-3]. We herein report a case of CSS who presented with distal asymmetrical polyneuropathy at the time of diagnosis, which rarely occurs.

CASE REPORT

A 26-year-old man was admitted to Srinagarind Hospital in February 1999 with symptoms of weakness and numbness of both hands and feet and

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arthralgia of the left knee for 2 weeks. Two months earlier, he had leg edema and 1 month later, he experienced fever and arthralgia over the right elbow. He had had late-onset asthma for the past 3 years.

On physical examination, he was a sthenic man, alert with a body temperature of 38.8°C and nonpitting edema of both feet. Muscle weakness was mainly distal and asymmetrical: right upper limb-proximal group grade 5/5, wrist flexor and extensor and small hand muscles grade 3/5; left upper limb-proximal group grade 5/5, wrist flexor and extensor and small hand muscles grade 0/5; right lower limb-proximal group grade 5/5, ankle dorsiflex and plantarflex grade 4/5; left lower limb-proximal group grade 5/5, ankle dorsiflex grade 2/5, ankle plantarflex grade 4/5. Deep tendon reflexes revealed generalized hyporeflexia. Sensory examination revealed glove and stocking type of pinprick sensory loss and hypoesthesia over the lateral aspect of the right upper arm and the right thigh. The propioceptive sensation was absent in all toes.

Certain groups of painless palpable purpura were detected at the medial aspect of both thighs, legs and dorsum of both feet. Other general and neurological examination were normal.

Complete blood count revealed hematocrit of 38 per cent; white blood cell count of 21,700 cells/mm^3 with 35 per cent polymorphonuclear cells, 14 per cent lymphocytes, 4 per cent monocytes and 45 per cent eosinophils. Erythrocyte sedimentation rate (ESR) was 50 mm/hr. Rheumatoid factor and antinuclear antibody (peripheral type) were positive. Blood urea nitrogen, creatinine, fasting blood glucose, electrolytes, VDRL, TPHA, anti-HIV, hemoculture, urine examination and chest X-ray were within normal limits. Electrodiagnostic study of peripheral nerves was consistent with polyneuropathy, predominantly axonopathy type. Skin biopsy of the left thigh revealed leukocytoclastic vasculitis with predominantly eosinophilic infiltration of the vessels and perivascular tissues. Palisading granulomas with central necrosis was not found in our specimen.

CSS was diagnosed and the patient was treated with prednisolone which resulted in a good response. His motor power gradually improved. On follow-up 1 year later, his motor power had returned to normal except power of the left wrist flexor grade 3/5, wrist extensor grade 4/5 and small hand muscles grade 3/5.

DISCUSSION

The clinical clues for the diagnosis of CSS include bronchial asthma, peripheral eosinophilia (>1500/mm^3 or >10 %), clinical manifestation consistent with systemic vasculitis and a histopathologic finding of vasculitis. Characteristic histopathologic findings are necrotizing arteritis or venulitis, eosinophilic infiltration and extravascular granulomas. Most patients have a history of asthma when CSS is diagnosed. However, a case of CSS without preexisting asthma has been reported[4]. Neurologic involvement affects about 60 to 70 per cent of patients [1,2]. Mononeuritis multiplex is the most common manifestation in the initial phase. During progression of the disease, distal symmetrical polyneuropathy usually develops. Patients with CSS are largely improved by corticosteroid treatment and, in severe cases, adjunction of immunosuppressive agents may be needed. Severe gastrointestinal tract or myocardial involvement is significantly associated with a poor clinical outcome[2].

In our patient, CSS was definitely diagnosed from clinical presentation with histological findings and distal asymmetrical polyneuropathy markedly improved by corticosteroid treatment. Early recognition and early treatment of this syndrome can prevent serious complications.

(Received for publication on February 16, 2000)

REFERENCES

1. Sehgal M, Swanson JW, DeRemee RA, Colby TV. Neurologic manifestations of Churg-Strauss syndrome. Mayo Clin Proc 1995; 70: 337-41.
2. Guillevin L, Cohen P, Gayraud M, Lhote F, Jarrousse B, Casassus P. Churg-Strauss syndrome. Clinical study and long-term follow-up of 96 patients. Medicine 1999; 78: 26-37.
3. Hattori N, Ichimura M, Nagamatsu M, et al. Clinicopathological features of Churg-Strauss syndrome-associated neuropathy. Brain 1999; 122: 427-39.
4. Chen KR, Ohata Y, Sakurai M, Nakayama H. Churg-Strauss syndrome: report of a case without preexisting asthma. J Dermatol 1992; 19: 40-7.

ประสาทส่วนปลายอักเสบแบบไม่สมดุลที่เกิดร่วมกับกลุ่มอาการเชอร์จ–สเตราสส์

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รายงานผู้ป่วยชายอายุ 26 ปี มาโรงพยาบาลด้วยอาการ ไข้ ขาบวม อ่อนแรงและชาของมือและเท้าทั้งสองข้าง และผื่นที่ผิวหนัง ผู้ป่วยมีประวัติเป็นโรคหอบหืดและปวดข้อร่วมด้วย การตรวจร่างกายทางระบบประสาทพบอาการที่เกิดจากความผิดปกติของเส้นประสาทหลายเส้นที่เป็นสองข้างไม่เท่ากัน การตรวจวินิจฉัยทางไฟฟ้าของเส้นประสาทเข้าได้กับความผิดปกติของเส้นประสาทหลายเส้น จุลพยาธิวิทยาของชิ้นเนื้อผิวหนังที่เป็นผื่น พบมีการอักเสบของหลอดเลือดร่วมกับมีเซลล์อีโอสิโนฟิลเด่น อาการทางระบบประสาทของผู้ป่วยดีขึ้นมากหลังจากการรักษาด้วยยาเพรดนิโสโลน

คำสำคัญ : ประสาทส่วนปลายอักเสบแบบไม่สมดุล, กลุ่มอาการเชอร์จ–สเตราสส์

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จดหมายเหตุทางแพทย์ ฯ 2544; 84: 907–909

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