Case Report

Guillain-Barre Syndrome in a patient with systemic vasculitis suggestive of Churg-Strauss Syndrome.

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A 48-year-old man presented with pins and needle-like sensations in the fingertips and the toes of one week duration, which progressed to involve both feet. He had an episode of severe low back pain without radiation. Since then his walking started to deteriorate and he found it difficult to ambulate, and walk up or down the stairs without assistance. Over the next few days, his lower back pain resolved but his walking difficulties continued worsening. He experienced abdominal cramps few days prior to admission. He denied any bladder dysfunction, neck pains or any recent trauma or fever.

Two days later, he developed a rash that affected his axillary region, back, abdomen, and inner thighs. He reported dry cough, which was increasingly progressive for the last few days. He denied diplopia, dysphagia or disequilibrium. He took Amitriptyline and B-complex for two days which then he discontinued for lack of perceived benefits. The patient's past history included hypertension since the age of 18 and Gout.

On admission he was afebrile, with stable vital signs. The clinical examination was significant for minimal weakness of the upper and lower limbs. Deep tendon reflexes were equal and symmetric except for mild hyporeflexia of the right-sided ankle jerk and absent left-sided ankle jerk. There was a distal sensory loss in a glove-stocking distribution. In addition, the patient displayed a macular rash involving the axilla, inner thigh, lower abdomen, and lower back.

EMG study was indicative of a primarily demyelinating polyneuropathy as evidenced by prolonged latencies and F waves with motor greater than sensory involvement and probable distal conduction block. The findings were consistent, though not diagnostic of acute idiopathic demyelinating polyneuropathy (AIDP).

Lumbar puncture revealed elevated protein at 1.24 g/L with no cells MRI of the brain with and without gadolinium did not reveal any abnormalities. MRI of the cervical spine revealed evidence of right-sided C6-C7 disc herniation without evidence of cord compression. His VC was 1.8 (64% predicted). Intravenous immunoglobin therapy for the presumptive diagnosis of Guillain Barre Syndrome (GBS) was initiated on day one, but the patient's condition did deteriorate and he suffered from progressive weakness, he became essentially quadriplegic and was areflexic.

The maximum strength in all extremities was no more than 1 or 2/5. His vital capacity dropped to 1.4 liters and he was transferred to the intensive care unit. Routine laboratory investigations revealed the following:

leukocytosis of 22.1 with 31% eosinophils (>6000), normal hemoglobin and platelets, CRP was elevated and the coagulation profile was normal. Serum IgE was >5000. Stool analysis was negative for ova or parasites. Tests were within normal range for renal function, CK, lactate, blood gases, ESR, Rheumatoid factor, but slightly elevated liver enzymes. Serology was negative for Mycoplasma pneumonia, cytomegalo-virus, Epstein-Barr virus, Rubella, B. Burgdroferi, Varicella, HBsAg, and anti-HCV. A Positive p-ANCA was detected, and was positive for anti-MPO. CT chest, abdomen, and pelvis showed slightly enlarged axillary, mediastinal, para-aortic and mesenteric lymph nodes with patchy shadowing in the lung and single pulmonary nodule (see figures 1 and 2). It also showed slight hepatosplenomegally.

Endoscopy was also performed and showed mild gastritis and ulcerative iliocecal valve. Campylobacter jejuni was negative. Peripheral blood smear showed significant Eosinophilia, but the eosinophils were mature, and per our hematologist did show the picture of hypereosinophillic syndrome. Transbronchial biopsy was performed and revealed evidence of vasculitis with Eosinophilia. (figure 3). Skin biopsy was nonspecific. At this time a coexistent P-ANCA associated vasculitis with neuropathy was considered, and pulse dose steroid was initiated.

The question was whether to consider more aggressive immunosuppressive therapies, keeping in mind the vasculitis component of the diagnosis. Ultimately, cyclophosphamide was initiated. Later on patient showed some improvement and started to move his upper extremities with 3/5 strength. His vital capacity improved to 1.85 liters. He remained areflexic but his mental status and cranial nerves were not involved. The patient now is dramatically improving and receiving intensive physiotherapy.

Discussion:

Our patient presented with acute fulminant neuropathy which showed characteristic features of GBS. This was strongly suggested by the rapid progression of symptoms over days and this was supported by the EMG study and the CSF albuminocytologic dissociation. However, a high evidence of vasculitis was essentially present. The patient history of skin rash, gastrointestinal complains, cough, Eosinophilia with high IgE, Positive p-ANCA and the histopathological findings of the transbronchial biopsy were altogether pointing towards vasculitis.

In spite of the fact that mononeuritis multiplex is more specific for the diagnosis of vasculitis neuropathy, some literatures showed some evidence that the pattern of nerve involvement may become more symmetric in later stage of the disease. Many alternative pathophysiological sequences can be proposed in our patient. One possibility, that patient was in the eosinophllic phase of vasculitis of Churg-Strauss Syndrome, an early latent form of the disease, which confused putting up the diagnosis of GBS which was, in turn, the primary cause of the whole neurological sequences. An alternative view that of a common pathological insult triggering both GBS and vasculitis and ultimately leading to the fulminant course of the disease.

In summary, by illustrating the complex cases involving acute polyneuropathy, we stress the importance of more time spent to

diagnose the disease. The diagnosis of GBS should not preclude other underlying complex pathological processes that could participate actively in the disease, or underestimating the addition of any other modalities of management.

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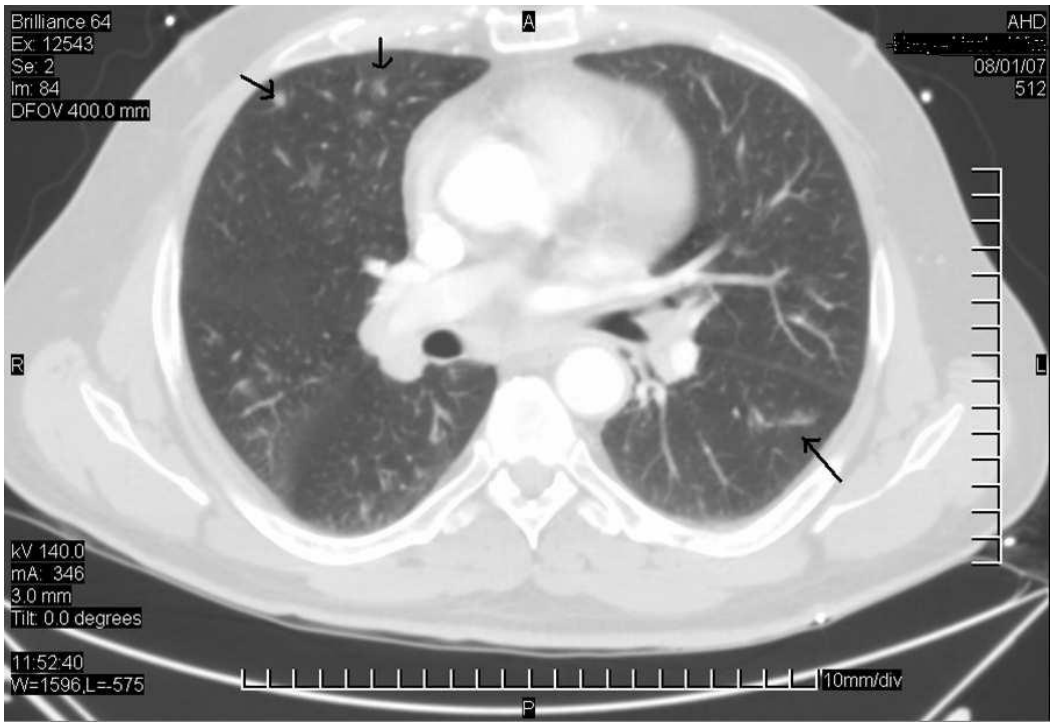

Figure 1: Chest CT scan showing patchy infiltrate in the RML, LUL and single pulmonary nodule in RML.

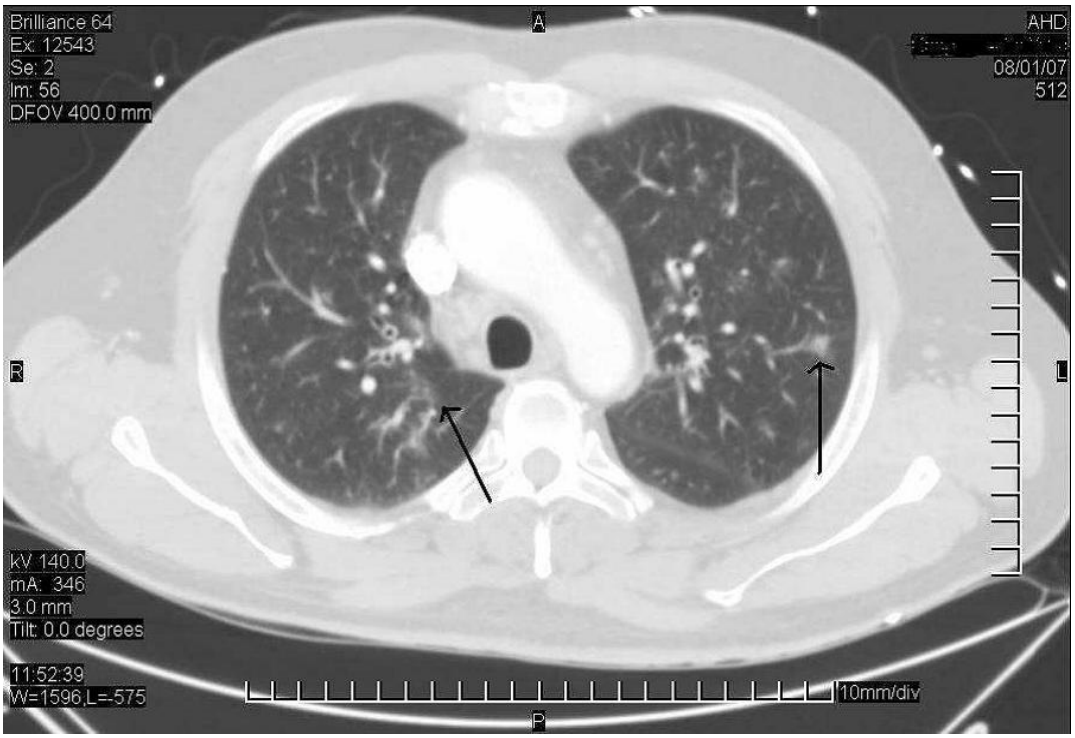

Figure 2: Chest CT scan showing patchy infiltrate (arrows) in the RUL, LUL.

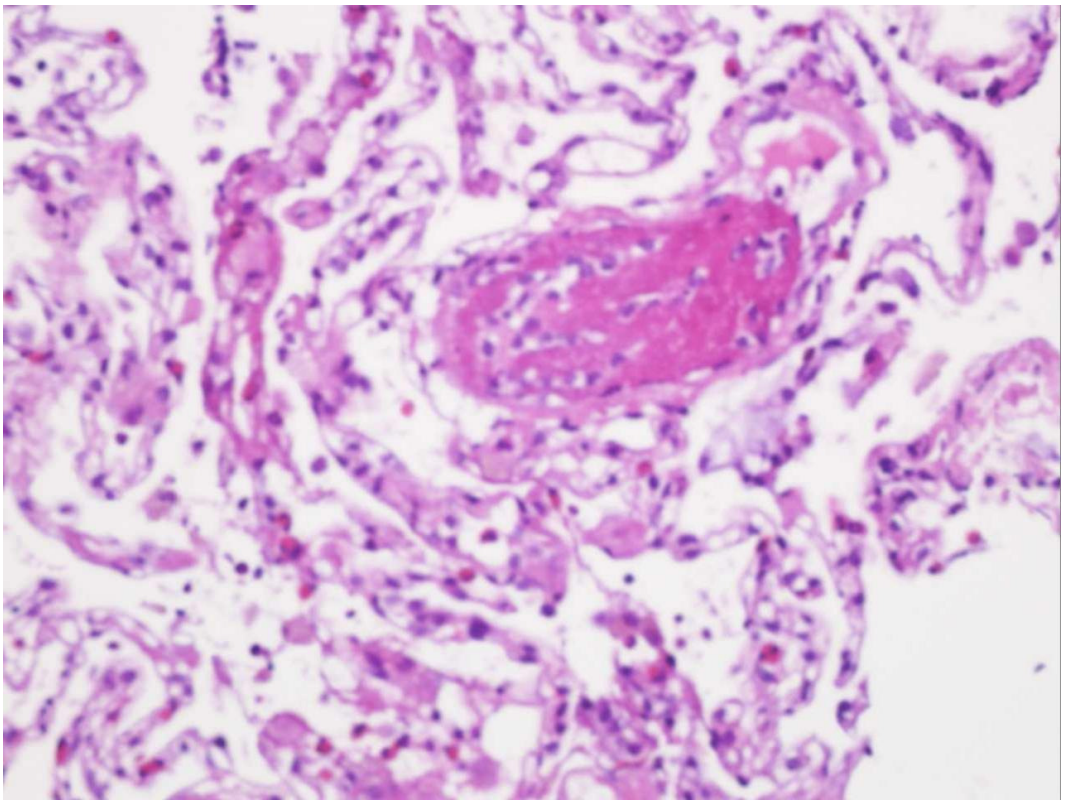

Figure 3: Focal vasculitis with eosinophils, compatible with Churg-Strauss syndrome.