

# An overlapping case of Bickerstaff brainstem encephalitis and acute motor axonal neuropathy variant of Guillain–Barré syndrome associated with systemic lupus erythematosus

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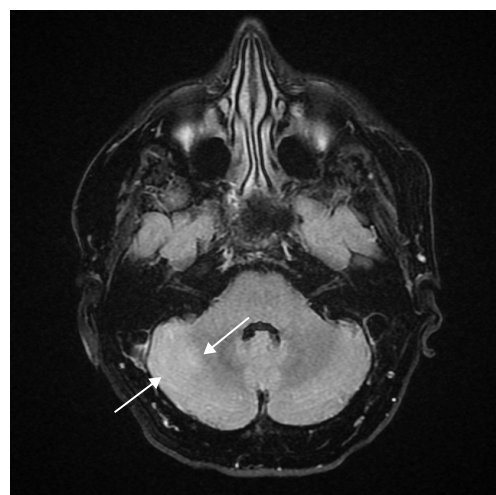
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Bickerstaff brainstem encephalitis (BBE) is a rare autoimmune disorder of the central nervous system. The clinical picture of BBE consists of ataxia, ophthalmoplegia, increased deep reflexes, and impaired consciousness.<sup>1</sup> It is often associated with the presence of anti-GQ1b antibodies.<sup>2</sup> Acute motor axonal neuropathy (AMAN) is a purely motor variant of Guillain–Barré syndrome.<sup>3</sup> This variant usually presents as progressive symmetrical weakening of the limbs with areflexia, often leading to respiratory failure. AMAN is associated with the presence of anti-GM1 ganglioside antibodies.<sup>4</sup> We present a case in which these 2 pathological syndromes coexisted with systemic lupus erythematosus (SLE).

A 41-year-old male patient with SLE experienced dizziness. On brain computed tomography, inflammatory lesions within the paranasal sinuses were visible. An antibiotic was applied and the patient was sent home. He returned to the hospital 2 days later because of worsening dizziness, ill feeling, fever, and vision disorders. There was a horizontal gaze nystagmus when looking to the sides, eye movement disorders, and increased deep reflexes on both sides, with a bilateral tendency of the Babinski sign. Brain magnetic resonance imaging (MRI) showed that the right cerebellar hemisphere had a blurred area with increased signal in T2 images and fluid-attenuated inversion recovery sequences (FIGURE 1). MRI perfusion of the brain revealed the features of small hyperemia in the right cerebellar hemisphere, which indicated inflammation. Methylprednisolone was introduced for treatment (1 cycle of pulse therapy administered at 1000 mg for 3 consecutive days). Despite the corticosteroid therapy, the general and neurological condition of the patient was

deteriorating. In the following days of hospitalization, the patient developed a full-blown cerebellar syndrome, consciousness disorders, and flaccid paralysis of the 4 limbs. Electromyography revealed axonal motor neuropathy, while electroencephalography showed numerous theta waves in all leads. The protein concentration in the cerebrospinal fluid was 99.4 mg/dl, associated with pleocytosis (60 cells/μl). The presence of anti-GM1 antibodies was confirmed, but not of anti-GQ1b antibodies. Bacterial and viral infections as well as paraneoplastic syndrome were excluded. Bickerstaff encephalitis with concurrent AMAN was identified.



**FIGURE 1** Magnetic resonance imaging in a 41-year-old patient, showing increased signal in the right cerebellar hemisphere (arrows) on a fluid-attenuated inversion recovery image

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In the treatment of the patient during the next 9 months, plasmapheresis (22 exchanges), cycles of intravenous immunoglobulin (6 cycles, 0.4 g/kg/d × 5 days), and rehabilitation were used alternately, but without any improvement in the patient's status. From the 10th month of hospitalization, the neurological condition of the patient slowly began to improve. There was a gradual return of deep reflexes in the upper limbs and knee reflexes, and the patient began to move the upper limbs. Intensive physical and speech rehabilitation was started. In the 11th month of hospitalization, the patient began to speak single words, follow instructions, and swallow. At 12 months, he was discharged home. A cerebellar syndrome with dominant ataxia of the torso and limbs as well as damage of both the upper and lower motor neurons were still observed. The patient remained in bed and required artificial ventilation.

This is the first case of a patient with SLE and overlapping BBE and AMAN reported in the available literature. Our case suggests that SLE, BBE, and AMAN can be part of a common immune spectrum.

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