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An overlapping case of Bickerstaff brainstem encephalitis and acute motor axonal neuropathy variant of Guillain-Barre syndrome associated with Systemic Lupus Erythematosus.

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Short title: BBE and AMAN variant of GBS syndrome associated with SLE.

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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 [1]. It is often associated with the presence of anti-GQ1b antibodies [2]. Acute motor axonal neuropathy (AMAN) is a purely motor variant of Guillain-Barre syndrome [3]. This variant is usually presented clinically in the form of a progressive symmetrical weakening of the limbs with areflexia, often leading to respiratory failure. AMAN is associated with the presence of anti-GM1 ganglioside antibodies [4]. Below we present a case in which these two pathological syndromes coexisted with systemic lupus erythematosus (SLE).

The 41-year-old patient with SLE experienced dizziness. In the brain computed tomography, inflammatory changes within the paranasal sinuses were visible. An antibiotic was applied and the patient was sent to home. The patient returned to the hospital two 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 Babinski sign. The magnetic resonance imaging (MRI) of the brain showed that the right cerebellar hemisphere had a blurred area with increased signal in T2-dependent images and FLAIR sequences (Fig.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 (one cycle of pulse therapy administered at 1000mg for three 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 four limbs. The electromyography (EMG) revealed axonal motor neuropathy, while electroencephalography (EEG) revealed numerous theta waves in all leads. The protein concentration in the cerebrospinal fluid was 99.4 mg/dL associated with pleocytosis (60 cells/ul). Anti-GM1 antibodies were found while anti-GQ1b weren’t found. Bacterial, viral infections and paraneoplastic
syndrome were excluded. Bickerstaff encephalitis with concurrent AMAN were identified. In the treatment of the patient during the next 9 months, plasmapheresis (22 exchanges), immunoglobulin cycles (6 cycles, 0.4g/kg/day x 5 days) and rehabilitation were used alternately but without any improvement to the patient’s status. From the 10th month of hospitalization, the neurological condition of the patient gradually began to improve. There was a gradual return of deep reflexes in the upper limbs and knee reflexes, the patient began to move the upper limbs. Intensive physical and speech therapy rehabilitation began. In the 11th month of hospitalization, the patient began to speak single words, follow instructions, and swallow. At 12 months the patient was discharged to home. A cerebellar syndrome with dominant ataxia of the torso and limbs and mixed damage of the upper and lower motor neuron 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 parts of a common immune spectrum.

REFERENCES


Fig 1. Magnetic resonance imaging in an 41-year-old patient showing increased signal in the right cerebellar hemisphere on fluid-attenuated inversion recovery (FLAIR) images.