Abstract

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Just in time: a rare case of AMSAN Guillain-Barre syndrome

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Background

Guillain-Barre syndrome is a rare autoimmune acute polyradiculoneuropathy, usually following an infection or other immune-stimulating event from 1 to 4 weeks. It usually presents with bilateral weakness in the distal lower limbs that gets progressively worse over days-to-weeks, leading sometimes to potentially life-threatening severity requiring mechanical ventilation. Different types of the syndrome have been described: acute inflammatory demyelinating polyneuropathy (AIDP, most common type), acute motor-sensory axonal neuropathy (AMSAN), and some other rare variants.

Case history

a 51 years old man was admitted at Emergency Room for abdominal pain and constipation. He was already hospitalized 3 days before for the same reason and discharged. At admission he complained hyposthenia and diffuse paraesthesia in the four limbs. Clinical evaluation showed impaired walking, ataxia, dysmetria, tremors in the upper limbs, and diffuses paraesthesia at the 4 limbs and complete abolition of osteon-tendon reflexes. Patient had a healthy lifestyle. However, he complained flu about 3 weeks ago. Head CT-scan excluded acute stroke and encefalus and spinal cord MRI excluded any other relevant disease of the CNS. Thus, Guillain-Barre syndrome was suspected and rachicentesis was performed: it showed high levels of albumin and a normal white blood cell count. Electromyography was performed, describing a polyradiculoneuropathy compatible with Guillain-Barrè syndrome AMSAN type. Prompt intravenous immunoglobulins 0.4 g/kg once a day for 5 days was administered and an overall progressive and slight improvement was observed. After 30 days was than discharged home.

Discussion

Guillain-Barre syndrome annual incidence is about 0.8-1.9 casese /100.000 persons in Europe. It typically presents with weakness at the extremity of lower limbs, then radiating proximally up to the upper limbs. It gets worse in 2-4 weeks with subsequent plateau that may last for several months or years before substantial improvement. Dysautonomia with urological or alvo dysfunctions may be associated. AMSAN–type has both weakness and sensory involvement, is more protracted and may have slower and less improvement. Attention should be paid to the most dangerous life-threatening

Symptoms

Respiratory failure, cardiac arithmia, dysphagia, adynamic ileus and hemodynamic instability. The 87% of patients report a total recovery or partial with minor deficits, as residual pain and fatigue, within 1-3 years of the onset. Mortality is about 3-7%. Timely start of therapy is the key for a better prognosis: within 2 weeks of the onset of symptoms if the patient needs help walking; otherwise, within 4 weeks (WHO interim guidance 2016). Fortunately, in our case diagnosis was made within a week after the onset and therapy was immediately started. No serious life-threatening symptoms and no complications related to therapy have been observed. Actually, he continues rehabilitation and everything gives hope for further improvement.