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Case report

Critical illness polyneuropathy or axonal Guillain-Barré syndrome triggered by subarachnoid haemorrhage?

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Abstract

Introduction

This article reports a case of the difficulty to differentiate between critical illness polyneuropathy and axonal Guillain–Barré syndrome when triggered by subarachnoid haemorrhage.

Case report

An 81-year-old man was admitted comatose (Glasgow coma scale score 4/15) after a subarachnoid haemorrhage. His neurological condition gradually improved with as best motor response (M4) withdrawal from pain at the four limbs. The patient developed early complications such as septicaemia and acute renal injury. After 3 weeks, a marked decrease of motor response (M1) was noted in the lower, and, to lesser extent, upper limbs. Deep tendon reflexes were abolished. The cerebrospinal fluid examination showed elevated protein level. After electrophysiological examination, the diagnosis of acute motor axonal neuropathy, a variant of Guillain–Barré syndrome, was discussed versus critical illness polyneuropathy. Specific therapy for Guillain–Barré syndrome could not be administered. No significant motor recovery was observed after 7 months.

Conclusion

The distinction between critical illness polyneuropathy and Guillain–Barré syndrome remains difficult in critically ill patients. It is not known if subarachnoid haemorrhage could be considered as a possible triggering factor for Guillain–Barré syndrome.

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Introduction

After subarachnoid haemorrhage (SAH), the patient's neurological condition may fluctuate over the first hospital days or weeks according to the magnitude of the bleeding and related complications. In patients with deeply altered consciousness, the testing of the best motor response may be challenging and deficits appear to be mainly due to central injury. The discrepancy between a progressive recovery of consciousness manifested as spontaneous eye opening or response to verbal command in the oro-facial territories and a persisting quadriplegia or quadriparesia should prompt further investigations to rule out peripheral injury. Numerous conditions have been associated with critical illness polyneuropathy (CIP) and myopathy and Guillain–Barré syndrome (GBS), including head trauma or intracerebral haemorrhage[1–4]. The potential role of subarachnoid bleeding is speculative, but the following observation illustrates the difficulties of the differential diagnosis between CIP and GBS when consciousness is impaired.

Case report

An 81-year-old man was admitted comatose in the emergency department. He had a medical history of arterial hypertension and type 2 diabetes mellitus; there was no evidence for a recent infection or vaccination. The initial Glasgow coma score (GCS) was 4/15 (E1V1M2) and intubation was required for mechanical ventilation in the intensive care unit (ICU). The diagnosis of SAH was made on the admission brain computed tomography (CT) (grade IV according to Fisher classification). No intracranial aneurysm could be demonstrated after contrast enhancement at CT and conventional angiography. The patient developed as an immediate complication a mild hydrocephalus, but the GCS gradually improved up to 10/15 (E4V1M6). However, the progression of hydrocephalus resulted in a neurological worsening (GCS ranging from 5 to 8). Despite ventricular drainage, the neurological condition remained unchanged. The best motor response was M4 in the four limbs. The patient developed a septicaemia due to *Staphylococcus aureus*, and a worsening of renal function was also noted. Haemodynamic instability was observed with alternating hypo- and hypertension. The repeated brain CT showed a regression of subarachnoid bleeding and hydrocephalus. After 3 weeks in the ICU, a marked decrease of motor response (M1) was noted in the lower, and, to lesser extent, upper limbs. The patient was able to open the eyes and the mouth on verbal command. Cranial nerve examination was normal. The deep tendon reflexes were abolished. The analysis of the cerebrospinal fluid (CSF) revealed elevated CSF protein level (122 mg/dl), with normal CSF cell counts. In the blood investigations, there was a polyclonal increase of immunoglobulins A and G; antibodies against myelin-associated glycoproteins were negative. Serum creatinine phosphokinase levels were elevated at the time of admission, but rapidly decreased. Routine serological investigations for common viral or bacterial infections were also negative. The differential

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diagnosis had to be made between GBS and CIP. The motor nerve conduction study was consistent with a severe axonopathy (absent distal compound muscle action potentials) and mild reduction of conduction velocities. Due to the presence of peripheral oedema at the four limbs, it was not possible to demonstrate a reduction of sensory nerve action potential amplitudes. On needle examination, motor unit recruitment was severely reduced in one territory and absent in the others; no denervation potentials were seen. No nerve or muscle biopsy was obtained. No specific therapy could be applied as plasma exchanges, and high-dose intravenous immunoglobulins were not tolerated (respectively due to marked hypotensive episodes and worsening of renal failure). After a prolonged ICU stay for more than 7 months, the patient remained quadriparetic and the weaning from mechanical ventilation was impossible; death was related to infectious complications. We concluded that the clinical course was more consistent with an axonal form of GBS.

Discussion

There are in the literature very few cases of GBS following either spontaneous or post-traumatic brain haemorrhage and the causal relationship between remains speculative[1–4]. A 29-year-old man developed a flaccid tetraparesia and respiratory failure 13 days following post-traumatic SAH[5]. High protein content (200 mg/ml) was found in the CSF, but this increased concentration could be easily explained by SAH. The electromyography was consistent with a demyelinating pattern. The patient experienced dysautonomic and infectious complications and died on day 19.

GBS is associated with cellular and humoral immune changes that could be eventually triggered by oxidative stress and oxidative damage after intracerebral haemorrhage, but this mechanism remains highly speculative[1,6].

In ICU patients, the differential diagnosis should certainly include CIP. Some authors suggest that there is a nosological overlap between acute motor axonal neuropathy, a variant of GBS, and CIP[7–9]. The distinction between the two entities cannot be easily made on clinical grounds. A marked albuminocytologic dissociation and autonomic instability are highly suggestive of GBS, but SAH may preclude the correct interpretation of CSF.

Electrophysiological studies of the pattern axonal GBS show a rapid fall in compound muscle action potentials and sensory nerve action potentials, with minimal evidence of demyelination. The initial needle EMG study lacks spontaneous activity. In CIP, there is a predominantly motor dysfunction. The morphological features in the nerve point to a primarily distal axonal degeneration of motor and sensory fibres.

Conclusion

The distinction between axonal forms of GBS and CIP remains difficult in critically ill patients. Patients with known risk factors for critical illness polyneuro(myo)pathy could still develop variants of GBS, with a different prognosis and treatment. In doubtful cases, electrophysiological investigations should be repeated, and the determination of antiganglioside antibodies could be helpful.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the editor-in-chief of this journal.

Abbreviations list

CIP, critical illness polyneuropathy; CSF, cerebrospinal fluid; CT, computed tomography; GBS, Guillain–Barré syndrome; GCS, Glasgow coma score; ICU, intensive care unit; SAH, subarachnoid haemorrhage

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