

Acute rhabdomyolysis associated with atypical Guillain-Barré syndrome

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Summary: We report a patient with atypical Guillain-Barré syndrome associated with acute rhabdomyolysis. Rhabdomyolysis may be the cause of elevation of creatine kinase sometimes seen in patients with Guillain-Barré syndrome.

Introduction

Mild elevation of creatine kinase (CK) is recognized in Guillain-Barré syndrome (GBS) but the cause is not known.¹ We report a case of GBS associated with marked elevation of CK and muscle biopsy features of acute rhabdomyolysis.

Case report

A 25 year old man developed anterior chest pain radiating to the inner aspect of both arms. The pain started suddenly and resolved over one hour. Three hours later, he developed weakness of his hands and legs which progressed over several hours until he was unable to walk. He had no sensory symptoms. There had been no recent illness or trauma, and his only past medical problem was childhood asthma.

On admission to hospital the next day he was mildly dehydrated, and had urinary retention requiring catheterization. General physical examination was otherwise normal. On examination of the nervous system, the cranial nerves were normal. In the arms, there was profound weakness of intrinsic hand muscles and finger flexion, with mild weakness of wrist flexion and finger extension. There was flaccid weakness of the legs, complete on the right but for a flicker of hip flexion, with moderate proximal and mild distal weakness on the left. Tendon reflexes were normal in the arms but absent in the legs. The left plantar was flexor and the right mute. There was a sensory level to pinprick at T2 with impairment of vibration and light touch appreciation below the right knee.

Serum electrolytes, calcium and phosphate were normal, but serum urea and creatinine were initially elevated at 42 mmol/l and 484 µmol/l respec-

tively, falling to normal levels over 10 days. Urinalysis was normal. Serum CK was markedly elevated at 10,150 IU/l on admission, returning to normal over 3 weeks. The CK-MB isoenzyme was not detected, and electrocardiogram and chest X-ray were normal. Antibody titres against a range of common viruses, legionella and mycoplasma, were not elevated. Antinuclear factor and urinary porphyrins were not detected.

Cerebrospinal fluid (CSF) was examined on the second and fourteenth days of illness, and protein content was 0.28 and 0.16 g/l, respectively. No cells were seen on either occasion, and immunology was normal. A full length myelogram and magnetic resonance imaging of the cervical cord were normal. Muscle biopsy showed foci of necrotic and oedematous muscle with no inflammatory infiltrate, consistent with acute rhabdomyolysis.

Neurophysiological examination was performed on the third day of illness. Motor conduction studies and distal motor latencies in the upper and lower limbs were normal, but F-wave responses were absent, suggesting exclusive involvement of proximal segments. Sensory abnormalities were found only in the most distal nerves (absent medial plantar response with intact sural, median and ulnar sensory potentials). Thermal threshold studies were normal, indicating intact small fibre function. Needle electromyographic (EMG) studies of the most affected muscles showed no evidence of spontaneous activity or denervation, and there were no myopathic changes.

The neurophysiological studies were repeated after 2 weeks, demonstrating the evolution of a severe demyelinating neuropathy affecting both proximal and distal segments of motor and sensory nerves in all four limbs. Needle EMG showed mild axonal involvement in proximal and distal muscles. Severe widespread axonal involvement was apparent on repeat studies after the eighth week of illness.

The patient underwent plasmapheresis with little initial improvement. Three months later there was some recovery of muscle power but the affected muscles in the arms remained weak and had wasted. By 10 months after the acute illness he could walk with a stick, and had full return of bladder function.

Discussion

This patient's illness was characterized by an areflexic paraparesis with a motor level at C7, sensory level at T2 and painless urinary retention. Paraparesis resembling a spinal cord lesion is a recognized though unusual variant of GBS.² Prolonged urinary dysfunction, the presence of a sensory level and failure of the CSF protein to rise after 2 weeks are other recognized variant features.³ The diagnosis of GBS was strongly supported by sequential neurophysiological studies. Proximal segment abnormalities, as in our patient, may be the only diagnostic feature in up to one quarter of GBS patients examined within the first week of illness.⁴ The second electrophysiological examination confirmed the widespread demyelinating neuropathy characteristic of GBS. The development of axonal involvement in the third examination may account for the patient's protracted and incomplete recovery.⁵

The transient impairment of renal function was probably due to a combination of mild dehydra-

tion and urinary retention, although myoglobinuria may have been a contributing factor. Membranous nephropathy resulting in nephrotic syndrome has been reported in association with GBS,⁶ but there was no evidence of such a process in this patient.

Modestly raised levels of CK (up to seven times the upper limit of normal) have been documented in the early stages of GBS, correlating with the occurrence of pain,¹ which was a presenting symptom in this patient, although, unusually, localized to the chest and arms. The cause of raised levels of CK in early GBS is uncertain. Muscle denervation can result in release of muscle enzymes.⁷ However, at the onset of this patient's illness when CK was elevated, small fibre function was intact, implying no axonal damage at this stage.⁵ The muscle pathology in this patient was typical of acute rhabdomyolysis, which was not found in muscle biopsies from patients with GBS in Ropper's series.¹ Hassin⁸ reported a case of GBS with inflammatory and degenerative changes in muscle, but his interpretation of the pathological features has been disputed.¹

Raised levels of CK can be a feature of the early course of GBS, and may cause diagnostic confusion with other conditions particularly if, as in this case, the clinical picture is not typical. The occurrence of rhabdomyolysis in this case suggests that it may be the pathological basis for elevation in CK levels. The mechanism remains open to speculation.

References

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