A Case of Acute Inflammatory Polyneuropathy with Atypical Features

Atipik Özellikler Gösteren Akut Enflamatuvar Polinöropatili Bir Olgu

Abstract

A case report with progressive weakness, bilateral facial paralysis and acute motor axonal polyneuropathy demonstrating electrodiagnostic evaluation will be presented. The condition was preceded by diarrhea, flu-like infection, abdominal pain and gastric tenderness; eventually, autonomic manifestation with episodes of visual hallucinations and disordered speech was added. Relation with *Campylobacter jejuni* infection, Lyme disease and porphyria are debated in differential diagnosis. Western-blot tests confirmed Lyme disease, and porphyrin specific test results were found positive.

Key Words: porphyria; acute inflammatory polyneuropathy; Guillain–Barré syndrome; neuroborreliosis

Özet

Bilateral fasiyal paralizi, ilerleyici kas güçsüzlüğü ve elektrofizyolojik değerlendirmede akut motor aksonal polinöropati bulguları gösteren bir olgu sunulmaktadır. Öncesinde karın ağrısı, ishal ve grip benzeri şikayetlere sonrasında otonom anormallikler, görsel halüsinasyon epizotları, konuşma bozukluğu eklendi. *Campylobacter jejuni* enfeksiyonu, Lyme hastalığı ve porfiri ayırıcı tanısı yapıldı. Western-blot testi Lyme hastalığı birlikteliğini doğruladı. Ek olarak porfirine spesifik testler pozitif bulundu.

Anahtar Kelimeler: porfiri; akut enflamatuvar polinöropati; Guillain–Barré syndrome; nöroborelyoz

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INTRODUCTION

Acute inflammatory polyneuropathy (AIP) or Guillain–Barré syndrome (GBS) is an immune-mediated neuropathy characterized by acute weakness with sensory symptoms, autonomic manifestations and albuminocytological dissociation in the cerebrospinal fluid (CSF) (1,2). In the etiology, gastroenteritis due to *C. jejuni* and flu infection were suspected among others. GBS-like manifestations can also be seen in Lyme disease (LD), porphyria, botulism, and diphtheria (3,4).

Neurological signs can be seen in 10-15% of patients with LD (neuroborreliosis) (5). Headache, meningismus, cranial nerve involvement, radiculitis/meningoradiculitis and neuropathy are most frequently seen manifestations of neuroborreliosis (6). The neuropathy of LD may present with AIP symptoms. (7,8).

Porphyrias are infrequently seen diseases arising from deficient enzymatic activities of HEM biosynthesis. Especially in acute intermittent and variegate types, neurological symptoms may occur. In acute intermittent porphyria, there are acute episodes of abdominal pain, tachycardia, extremity and muscle pain, sensory deficit, and weakness of proximal muscles, being more prominent in the upper extremities. In the variegate type, abdominal pain, tachycardia, constipation, vomiting, hypertension, neuropathy, confusion, psychiatric symptoms, bulbar paralysis and dysuria are seen. Hyponatremia can also be seen in both types (9-11).

Here we present a case with extremity weakness, facial diplegia, abdominal pain, autonomic dysfunctions, dysarthria and delirium. LD test and porphyrin-specific test results were positive.

CASE REPORT

A 53-year-old male patient was admitted to our emergency clinic with complaints of widespread weakness and numbness. He was a farmer and had no known disease. One week previously, the patient had diarrhea and flu infection, and four days later he developed severe abdominal pain. He had undergone endoscopy and colonoscopy in another hospital and a diagnosis of gas entrapment was made. There was a tick bite history 3 years previously. In our examination, he was agitated, his voice was hoarse and trembling, and he had shortness of breath. Facial diplegia was found. There was weakness in all extremities (3/5), more prominent in the right arm (1-2/5). Deep tendon reflexes could not be elicited. His blood examination was normal except Na (128 mmol/L) and CRP (5.51 mg/L). Electromyography revealed markedly decreased amplitudes of motor nerve responses in the lower extremities and mild to moderately delayed distal latencies. Conduction block was not detected. Sensory responses were normal. F responses could be elicited with some delay. These findings made us think of a mainly axonal and motor neuropathy. In his lumbar puncture, CSF pressure was normal, the fluid was clear and devoid of any cells, its protein and albumin concentrations were 45.3 mg/dL and 28.8 mg/dL, respectively. CSF cultures were negative. With ELISA, IgG antibody was positive, IgM antibody was negative for LD. Blood and CSF Western-blot tests showed IgG positivity. *C. jejuni* could not be isolated from the stool culture. After two days of Ceftriaxone treatment (2 g. BID), weakness became more prominent in the right upper and lower extremities. His facial diplegia deteriorated, sweating attacks and constipation worsened. Eventually, his orientation became worse, visual hallucinations and slurred speech were added. EEGs demonstrated widespread disorganisation. In his cranial MRI on T2A, bilateral multiple hyperintense signals on the posterior crura of the internal capsule and cerebral peduncles were detected (figure 1,2). IVIG treatment at a total dose of 2g/kg was given. In his 24-hour urine sample, Coproporphyrin I, coproporphyrin III, and total porphyrins were 92.04 µg/dL (normal range 0-25), 80 µg/dL (0-75), and 196.70 µg/dL (0-150), respectively. For definitive diagnosis, genetic confirmation was required and the patient was referred to a medical genetic consultation for testing PBGD gene activity. After 6 weeks, neuropsychiatric features quickly declined, and he did not need mechanical ventilation. His arm strengths were still about 1-2/5, his legs were at

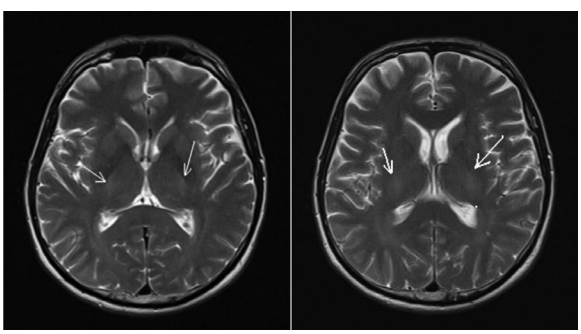

Figure 1: Cranial MRI. T2-weighted axial sections, hyperintense signals, on the posterior crura of the internal capsule bilaterally and cerebral peduncles.

(2-3/5). He was transferred to the Physical Treatment and Rehabilitation clinic.

DISCUSSION

In our case, LD and porphyria diagnoses were suggested because of acute motor neuropathy (AMN) with abdominal pain, autonomic dysfunction, and neuropsychiatric manifestations (12,13). Facial diplegia can be seen in both of these diseases, too (14). Because of preceding abdominal pain and diarrhea, *C. jejuni* infection-triggered acute motor axonal neuropathy (AMAN) was also another possibility.

In the literature, hallucinations, sleep disorders, depressive symptoms, and speech disorders have been described both in LD and porphyria (9,15). Hyponatremia, which was seen in our case as well, was thought to be responsible for neuropsychiatric findings both in LD and porphyria, secondary to inappropriate ADH syndrome in two reports (9,16). IVIG therapy may also cause hyponatremia. Since neuropsychiatric manifestations started before initiation of IVIG therapy, we evaluated these findings as being related to the disease itself.

In acute porphyria, characteristically upper extremities and proximal muscle groups are affected at the onset, and typical albuminocytologic dissociation seen in GBS is not expected (11), as was the case in our patient. CSF protein concentration was mildly increased; however, lymphopleocytosis was not detected. These findings with elevated urine coproporphyrin level made us get closer to the diagnosis of porphyria, but could not eliminate LD.

In porphyria, EMG findings have been described as signs of motor axonal neuropathy without conduction block or marked slowing of conduction velocity, which was compatible with our case (11,17). LD can have different neuropathic presentations, and GBS/GBS-like syndrome has also been reported with LD (18)

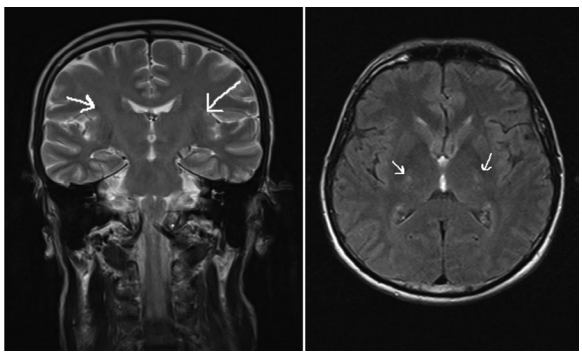

Figure 2: Cranial MRI. T2-weighted coronal and T2-FLAIR axial sections, hyperintense signals, on the posterior crura of the internal capsule bilaterally and cerebral peduncles

On the 2nd day of ceftriaxone treatment, the patient's confusion deteriorated and episodes of visual hallucinations were increased. These manifestations could be due to the fragmentation of bacteria with antibiotherapy with resultant aggravation of symptoms, like those occuring in spirochete infections (Jarisch-Herxheimer reaction). This reaction has also been reported in LD and can be observed after treatments with amoxicillin and doxycycline (19-21).

In the cranial MR of our patient, on T2A-weighed sequences, increased signal intensity in the corticospinal tract was detected, which is a specific finding of ALS (22). Such an MR finding in LD or porphyria has not been encountered in previous reports. However, there are some cases of AMNs with central demyelination. It was reported that central motor conduction time has been found significantly delayed in three GBS-diagnosed patients with hyperreflexia (23,24). In our case, after follow up for one year, hyperreflexia did not develop, hyperintesity on the posterior crura of the capsula interna decreased in control MR imaging.

Although there was AMN predominantly in the arms, as well as abdominal pain and neuropsychiatric manifestations, we could not establish a definitive diagnosis of porphyria because of inconclusive history, lack of darkening of urine and negative results in genetic tests.

The AMAN variant of GBS is characterized by pure motor involvement, frequently with previous *C. jejuni* infection. The isolation rate of *C. jejuni* from stool culture of these patients ranges from 8% to 50% for 1-3 weeks after diarrhea (25).

Lately, a few cases of changed mental status due to PRES (posterior reversible encephalopathy syndrome) with GBS or hyponatremia have been reported. Many of them had developed after IVIG therapy (26). Hyponatremia has been reported within a range of 26 to 31% of GBS patients. The precise pathogenetic mechanism of hyponatremia in GBS is uncertain. As data about rare findings of GBS have suggested, solely GBS itself may explain all of the features of our patient. Reports about GBS with *C. jejuni* is also supporting this possibility (27).

Although central involvement and change of the sensorium with GBS have not yet been included in the

absolute criteria, recently, they have been approved for the Bickerstaff encephalitis variant of GBS. Multiple T2 hyperintensities on the posterior leg of the capsula interna and the acute confusional status of our case may give some support for this diagnosis (28,29).

In conclusion, we could not reach an exact diagnosis because of limited laboratory support. However, some rare findings seen with GBS, LD, and porphyria should be kept in mind. Bickerstaff encephalitis can be another possibility if central involvement and/or change in sensorium occur.

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