Guillain-Barré Polyneuropathy Associated with Mediterranean Spotted Fever: Case Report

In Bulgaria, Rickettsia conorii, the etiologic agent of Mediterranean spotted fever (MSF), is transmitted by the dog tick Rhipicephalus sanguineus. The disease is associated with specific symptomatic signs, such as fever, headache, fatigue, and muscle and joint aches. One week before the onset of symptoms, he had had a tick bite. Four days after the onset of fever, a copious truncal rash appeared, and he was admitted to the hospital. His general condition was disabled, and he was febrile and weary. He had red conjunctivae and oropharyngeal mucosa and a dry coated tongue. A generalized papular rash extended from his trunk to the palms and soles. A skin lesion—tache noire—was evident on the inner side of the left thigh.

Laboratory evaluations showed leukocytosis with a shift to the left and an erythrocyte sedimentation rate of 50 mm/h. Neurological examination did not reveal abnormalities. He was treated with doxycycline (100 mg twice daily) for 7 days. The disease quickly responded to treatment: within 3 days, the fever abated, and the patient was discharged in good general and neurological condition.

On the next day, he was readmitted to the hospital because of severe back pain that spread to the flanks. A tingling sensation in the back, weakness of the lower limbs, and numbness of the fingers and toes appeared subsequently. The weakness increased quickly, and 5 days later, the patient was not able to walk without assistance. Neurological examination revealed latent paralysis, hypotonia, and areflexia of the arms. The lower limbs were flaccid with distal paresthesias. Within 1 day, quadriplegia developed, and the patient became bedridden. He was transferred to the intensive care ward.

An approximately symmetrical limb paralysis and right facial palsy were also noted. Deglutition was only merely impaired. The heart rate was regular and frequent (108 beats/min). CSF analysis showed an elevated protein concentration (0.9 g/L) without cellular response. Spinal CT did not show evidence of cord compression. After a total of 8 days, his condition stopped deteriorating, and a plateau phase lasted for 2 weeks. During the whole period of neurological impairment, the patient remained afebrile and conscious, and laboratory tests did not show any significant deviation in findings.

Electromyography results were consistent with the demyelination process of GBS, except for a single peculiarity in some axonal degeneration of the distal nerve. After the plateau phase, his condition progressively improved, leading to complete resolution of paralysis of the upper limbs and Bell’s palsy. About 2 months after the onset of GBS, the patient was almost entirely recovered; he was discharged with slight leg hyperesthesia. Ten months later, he had normal motor activity as well as normal electromyography findings.

Intravenous γ-globulin (2.5 g/d for 3 days) was added to therapy during the burst of neurological symptoms. Corticosteroid treatment was not used [2]. MSF was verified by indirect immunofluorescent antibody testing at a reference rickettsial laboratory. Three serum samples were collected 4, 12, and 28 days after the onset of MSF; titers of antibody to R. conorii were 1:20, 1:80, and >1:640, respectively.

In the available literature, we found only one case of R. conorii infection associated with polyradiculoneuritis that was reported in 1968 [3]; pleocytosis in this case was not consistent with GBS, and symptoms of MSF were not previously manifested. A case of GBS associated with Rocky Mountain spotted fever has been reported in the United States [4].

Our case was the only one in 300 patients with MSF who were observed during a 4-year period [1]. Our patient’s polyneuropathy satisfies the criteria of Asbury and Cornblath [5] for GBS. Electromyography findings were typical of those of GBS except for a single peculiarity: low-level expression of axonal degeneration. This finding is not strange because a variant of GBS (called axonal GBS) has been described in which rapid almost complete paralysis occurs [2].

A T lymphocyte mechanism is currently proposed for the inflammation in GBS. Activated by recent infection, T cells are sensitized to P-2, a major peripheral nerve myelin antigen. An early antibody attack on myelin is also possible [2]. This hypothesis concerns the molecular mimicry and is supported by the fact that approximately two-thirds of cases of GBS follow infectious episodes. However, we did not prove antecedent viral or bacterial infections in our patient. The disease had a benign self-limiting course. No relapse was observed.

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References