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ZIKA VIRUS-ASSOCIATED GUILLAIN-BARRÉ SYNDROME VARIANT IN HAITI

Zika virus is a single-stranded RNA virus (genus *Flavivirus*) transmitted by the *Aedes* mosquito and through sexual contact with infected individuals.^{1,2} Zika virus infection may be asymptomatic or may cause fever, rash, joint pain, conjunctivitis, myalgias, and headache.¹ Initially endemic to Africa and Asia, outbreaks in the Pacific Islands occurred in 2007 and 2013, and Central America, South America, and the Caribbean islands are currently in the midst of an epidemic. In both prior and current outbreaks of Zika,¹ an increased incidence of Guillain-Barré syndrome (GBS) has been reported.

We report a patient from Haiti who presented with the GBS variant facial diplegia with acral paresthesias and subsequently developed the features of Miller Fisher syndrome.

Case report. A 35-year-old man presented to his local hospital in Haiti in early January with acute onset of bilateral weakness and the sensation of “electrical currents” in his hands and feet. He reported headache, fever, nasal congestion, and eye pain several days before presentation, which had resolved at the time of onset of his neurologic symptoms. On examination, he had bilateral lower motor neuron-pattern facial weakness, generalized areflexia, and a mildly ataxic gait. A diagnosis of the facial diplegia with acral paresthesias variant of GBS^{3,4} was made.

The patient decided to pursue care in the United States. Several days after onset of bilateral paresis, he developed bilateral complete external ophthalmoplegia and mild upper extremity ataxia, and worsening gait ataxia in addition to the above-noted findings, consistent with Miller Fisher syndrome. He maintained a normal level of consciousness and normal strength. CSF analysis demonstrated protein 114 mg/dL, glucose 75 mg/dL, and <1 white blood cell/ μ L. MRI of the brain was normal. Rapid plasma reagin and HIV were negative. Serum and CSF Zika virus immunoglobulin M (IgM) were positive by both ELISA and plaque reduction neutralization test (1:10,240 in serum; 1:64 in CSF; Centers for Disease Control Laboratory). The patient was treated with IVIg (2 g/kg over 5

days). Serum anti-ganglioside antibody levels (GM1, GM2, GD1a, GD1b, GQ1b antibodies) sent after treatment with IVIg were within the normal range (Mayo Medical Laboratories).

At discharge, the patient had marked improvement in his ataxia and was able to walk with a cane. Three weeks after discharge, he had near complete resolution of ophthalmoplegia and return of all tendon reflexes, but he had minimal improvement in his bifacial weakness and continued to require a cane because of residual sensory gait ataxia.

Discussion. An increase in GBS incidence has been reported in Brazil, Colombia, El Salvador, Suriname, and Venezuela in the current Zika epidemic,¹ and evidence for the association of GBS with Zika infection has been reported from the 2013–2014 Zika epidemic in French Polynesia.⁵ It remains unknown whether an increase in GBS incidence in the setting of Zika epidemics is related to the widespread presence of a nonspecific infectious trigger of GBS, whether there is specific molecular mimicry to antigens presented by the Zika virus that leads to GBS, or whether there may be a direct effect of Zika infection on peripheral nerves.^{5,6}

In many Zika-endemic regions, access to neurologists may be limited. Although typical GBS is generally easily recognized by nonneurologists, variants such as facial diplegia with acral paresthesias and Miller Fisher syndrome may not be readily identified as GBS. Surveillance efforts⁷ must therefore emphasize the potential variety of GBS presentations in order to define the true incidence of GBS in Zika-affected regions, characterize the association between Zika and GBS, and determine whether there is a unique molecular mechanism of GBS in patients with Zika infection.

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Zika virus-associated Guillain-Barré syndrome variant in Haiti (see p. 336)

This podcast begins and closes with Dr. Robert Gross, Editor-in-Chief, briefly discussing highlighted articles from the July 19, 2016, issue of *Neurology*. In the second segment, Dr. Stephen Donahue talks with Dr. Aaron Berkowitz about his paper on Zika virus-associated Guillain-Barré syndrome variant in Haiti. Dr. Ilena George then reads the e-Pearl of the week about juvenile Huntington disease. Dr. Alberto Espay focuses his interview with Dr. Alireza Atri on his clinical trials plenary session at the AAN

Annual Meeting with regard to 5-HT₆ antagonist as an adjunctive therapy to cholinesterase inhibitors in patients with mild-to-moderate Alzheimer disease.

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