Cranial Nerve Enhancement in the Guillain-Barré Syndrome

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Summary: We report a case of Guillain-Barré syndrome with enhancement of multiple cranial nerves seen with postcontrast MR imaging. Clinical symptoms and electrodiagnostic studies reflected abnormalities of some but not all of the enhancing cranial nerves.

Index terms: Nerves, cranial; Spinal cord

The Guillain-Barré syndrome is an acute, demyelinating polyneuropathy of unknown cause (1, 2). Patients present with a progressive motor neuropathy, elevated cerebrospinal fluid protein, and usually diminished motor nerve conduction on electrodiagnostic studies. Clinical involvement of cranial nerves is common, manifested as opthalmoplegia and weakness of the facial and oropharyngeal musculature. A recent report documented bilateral facial nerve enhancement on magnetic resonance (MR) imaging (3). We report an additional case of Guillain-Barré syndrome with multiple cranial nerve enhancement seen on gadolinium-enhanced MR imaging.

Case Report

A 50-year-old woman presented with paresthesias and unilateral lower extremity weakness 1 week after a mild upper respiratory tract infection. On examination, she was afebrile, with mild right facial and lower extremity weakness. Her deep tendon reflexes were absent. Cerebrospinal fluid examination revealed a normal cell count and a protein concentration of 74 mg/dL; cultures for bacteria and fungi were negative. Chest roentgenograms, antinuclear antibodies, and Lyme titers were normal. Electrodiagnostic studies showed increased peroneal and tibial F-wave latencies and evidence of demyelination of both facial nerves proximal to the styloid-mastoid foramen, worse on the right side.

Three days into her hospital course, dysphagia developed, with progressive weakness of both sides of the face and lower extremities. On MR, enhancement after administration of gadolinium (0.1 mmol/kg) was seen in the internal auditory canal, geniculate ganglion, and tympanic portions of the right facial nerve (Fig 1A and B); in the cisternal segments of cranial nerves six bilaterally (Fig 1C and D), and in the right cranial nerve X and XI complex within the jugular foramen (Fig 1E and F). The patient underwent plasmapheresis, with near complete resolution of symptoms by 8 weeks. A follow-up MR was not obtained.

Discussion

The Guillain-Barré syndrome is an acute polyneuropathy characterized by limb weakness and mild sensory signs (1, 2, 4, 5). In approximately two thirds of cases, a mild respiratory or gastrointestinal tract infection precedes the onset of symptoms (2). Surgical procedures, lymphoma, and systemic lupus erythematosus are other predisposing factors (2). Cerebro-spinal fluid analysis typically reveals elevated protein concentrations without pleocytosis (1, 2). Clinical involvement of cranial nerves is common (1). Facial nerves are the most frequently affected, in approximately 50% of patients; compromise of the bulbar and oculomotor nerves also can be seen (1, 2). The majority of patients recover within a few weeks to months, with maximal paralysis usually reached by 4 weeks (1, 2). Early plasmapheresis often improves clinical response (4, 5). Intensive-care monitoring allows early treatment of cardiopulmonary complications.

The cause of the Guillain-Barré syndrome is unknown, with both humoral and cellular autoimmune mechanisms implicated (6, 7). In some patients, IgM antibodies to myelin glycolipids or gangliosides have been documented (7); other patients have evidence of T-cell hypersensitivity (2). Pathologically, there is an inflammatory infiltrate of mononuclear cells that surrounds small, endoneural vessels, and adjacent segmental demyelination (2).

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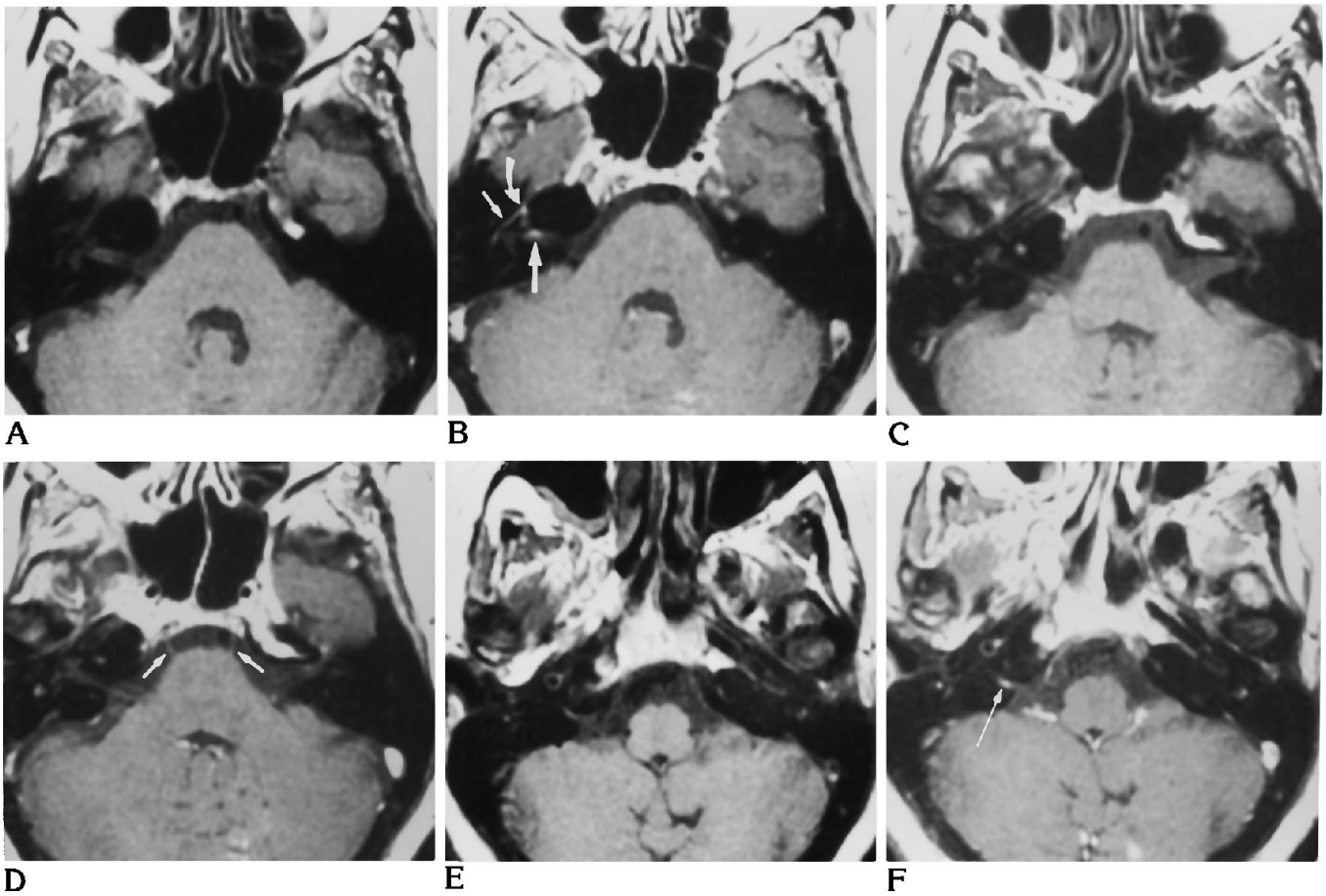

Fig 1. Cranial nerve enhancement in Guillain-Barré syndrome: Axial MR (500/20/2 [repetition time/echo time/excitations]) before (*A*, *C*, and *E*) and after (*B*, *D*, and *F*) single-dose gadolinium (0.1 mmol/kg) administration.

A and *B*, At level of internal auditory canal. Although enhancement of the proximal, greater superficial petrosal nerve and geniculate ganglion (*curved arrow, B*) and tympanic portions (*short, straight arrow, B*) of the facial nerve can be normal, enhancement within the internal auditory canal (*large, straight arrow, B*) indicates disease.

C and *D*, At level of pons. *Straight arrows* in D demonstrate enhancement of the cisternal portion of both cranial nerves 6.

E and *F*, At level of jugular fossa. After gadolinium administration, the cranial nerve X–XI complex enhances on the right (*arrow, F*).

The mechanism of abnormal enhancement of the cranial nerves in patients with Guillain-Barré syndrome is not entirely understood. There is evidence of a blood–peripheral nerve barrier as shown by failure of Trypan blue, a marker of an intact blood-brain barrier, to stain peripheral cranial nerves (8). Disruption of this blood-nerve barrier by the inflammatory infiltrate, analgous to enhancement in brain parenchyma after breakdown of the blood-brain barrier, therefore may cause enhancement. Alternatively, perineural structures, such as a vascular plexus enlarged from surrounding inflammation, could produce a local increase in gadolinium, and thus result in enhancement (9).

In this patient, enhancement of right cranial nerve VII and the cranial nerve X–XI complex on the right correlated with clinical findings of right facial and oropharyngeal musculature weakness and electrodiagnostic evidence of demyelination. Electrodiagnostic studies also indicated evidence of demyelination of cranial nerve VII on the left; however, we did not see enhancement. Conversely, both sixth cranial nerves enhanced, but no clinical evidence of demyelination was found. A larger number of patients must be studied—comparing gadolinium-enhanced MR, clinical exam, and electrodiagnostic studies—to determine how accurately gadolinium enhancement indicates cranial nerve involvement in the Guillain-Barré syndrome.

In conclusion, the Guillain-Barré syndrome should be considered in the differential diagnosis of cranial nerve enhancement, which includes infections (10–16), neoplasms (primary

or secondary) (15–22), granulomatous diseases (15, 17, 23), multiple sclerosis (24), vasculitic neuropathies such as diabetes mellitus (25), and unknown causes (26).

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