

Bilateral Radiation-Induced Hypoglossal Nerve Palsy Responsive to Steroid Treatment

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Dear Editor,

A 63-year-old woman reported a 2-month history of neck pain followed by progressive deterioration of speech and swallowing and a weight loss of 3 kilograms. Six years ago, she had received radiotherapy for a lymphoepithelial rhinopharyngeal carcinoma. A neurological examination showed dysarthria, dysphagia, impaired tongue protrusion, and amyotrophy on the left side. Blood screening for autoantibodies (antinuclear antibodies, extractable nuclear antibodies, anti-neutrophil cytoplasmic antibodies, and anti-DNA), Lyme-disease serology, angiotensin-converting enzyme, anti-acetylcholine-receptor antibodies, and HIV produced negative findings. A cerebrospinal fluid (CSF) examination showed normal protein levels, the absence of malignant cells, and negativity for an immunoelectrofocusing reaction. A CSF search for viral genomes by PCR excluded active infections with herpes simplex virus types 1 and 2, varicella zoster virus, Epstein-Barr virus, and cytomegalovirus. An echo color Doppler study of the extracranial vessels produced normal findings. Magnetic resonance imaging of the brain, nasopharynx, and neck with gadolinium enhancement excluded locoregional tumor relapse and meningeal carcinomatosis. The findings of a sensorimotor nerve conduction study, F-wave examination, and high- and low-frequency repetitive nerve stimulation were normal. In contrast, needle electromyography (EMG) disclosed active denervation of the tongue muscles (abundant fibrillations and positive sharp waves at rest in both genioglossus muscles with the absence of voluntary activity). Her motor evoked potentials were normal. A videofluoroscopic evaluation revealed severe dysphagia with a penetration-aspiration scale (PAS) score of 6 and a pooling score of 10. Percutaneous endoscopic gastrostomy was performed in order to assure adequate feeding and avoid the risk of aspiration.

A putative diagnosis of postradiation bilateral hypoglossal palsy was made, and treatment with a steroid (1 mg/kg prednisone) was started and then slowly tapered. At the 7-month follow-up the patient reported improvements of speech and swallowing, while the videofluoroscopic re-evaluation confirmed the improvement of dysphagia (PAS score of 4 and pooling score of 8), and so the patient was allowed to consume semisolid food orally. An EMG evaluation disclosed the absence of spontaneous activity and chronic neurogenic changes associated with diffuse phenomena of collateral reinnervation in the genioglossus muscles, confirming the functional recovery of hypoglossal nerves.

The differential diagnosis of bilateral hypoglossal palsy includes metastatic disease at the base of the skull, motor neuron disease, dissection of extracranial vessels, meningeal carcinomatosis, multiple mononeuritis of different etiologies (infective, vasculitic, and immune-mediated), and tumoral nerve infiltration. All of these conditions were ruled out in the present case. Cranial neuropathies are uncommon but well-recognized complications after head and neck radiotherapy, with an incidence ranging from 5% to 10%.^{1,2} The average latency period is reportedly 5 to 7 years.^{2,3} The hypoglossal nerve is the most frequently involved, often bi-

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laterally.^{1,2} This nerve exits through the hypoglossal canal, runs along the carotid space, and enters the tongue, and hence it is exposed over a long tract, which makes it at a higher risk than other cranial nerves to damage from high-dose irradiation. The pathogenesis of postradiation nerve palsy is controversial, with some authors suggesting compression of the nerve along its passage through the neck due to radiation-induced fibrosis, and other hypothesizing that the radiation-induced damage of small blood vessels supplying the nerve could lead to neural ischemia, demyelination, and fibrosis.³ Radiation-induced cranial neuropathies (RICN) are usually irreversible with no specific treatments being available, which causes a significant burden to the quality of life.^{1,2} A response to high-dose intravenous steroid was recently reported in a patient with long-standing RICN.⁴ In our case, treatment with an oral steroid led to the amelioration of symptoms and clinical picture. Moreover, EMG disclosed re-innervation phenomena suggestive of the functional recovery of the hypoglossal nerves. The response to steroid treatment observed in our patient could suggest alternative diagnoses, such as vasculitic neuropathy, chronic inflammatory demyelinating polyneuropathy, or cranial neuropathy associated with sarcoidosis. However, these conditions were ruled out by blood tests, a CSF examination, and neurophysiological investigations.

The improvement observed in the present case and the previously reported case question the historical view that RICN are irreversible and suggest that an underlying modifiable inflammatory component has a pathogenetic role in some cases. Steroids should therefore be considered an option treatment for patients with RICN in clinical practice. However, controlled clinical trials are warranted to confirm their efficacy and the optimal dose strength and route of administration.

Conflicts of Interest

The authors have no financial conflicts of interest.

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