Neuritis ossificans of a cranial nerve

Case report

Craig M. Kemper, M.D., Julio C. Rojas, M.D., Ph.D., and Steven Bauseman, M.D.

1Seton Brain and Spine Institute; 2Department of Psychology, University of Texas at Austin; and 3Department of Pathology, University of Texas Medical Branch, Austin Campus, Austin, Texas

The authors report the case of intracranial neuritis ossificans presenting as chronic accessory neuropathy. Neuritis ossificans is a rare reactive nerve disease that has been reported to affect systemic peripheral nerves. To the best of the authors’ knowledge, this is the first documented case of neuritis ossificans observed in a cranial nerve. The lesion was revealed on imaging studies and appeared, intraoperatively, as a nonossous intradural lesion with significant calcification of rootlets of the lower cranial nerves. Microscopically, the lesion featured zonal heterotopic calcification typical of neuritis ossificans. Although it is a rare entity, neuritis ossificans can be considered in the differential diagnosis of lower cranial nerve neuropathy. (DOI: 10.3171/2010.5.JNS091931)

Key Words • heterotopic ossification • intracranial • accessory • neuropathy • neuritis ossificans

Neuritis ossificans is a form of reactive heterotopic calcification that affects a segment of a nerve and that is usually confined to the epineurium. It is a rare entity that has been reported to affect peripheral nerves including the sciatic nerve, common peroneal nerve, ulnar nerve, median nerve, saphenous nerve, and tibial nerve, producing clinically meaningful sensory-motor deficits. Neuritis ossificans is believed to be secondary to trauma, given its pathological resemblance to myositis ossificans, although this etiological relationship remains unclear. The treatment of neuritis ossificans has been resection to prevent further clinical deterioration, and a nerve-sparing procedure if minimal fiber loss secondary to compression is present. No previous reports of neuritis ossificans affecting any cranial nerve exist in the literature. This report describes a case of histologically confirmed neuritis ossificans of the accessory nerve presenting as accessory neuropathy. The imaging features of this lesion are also described for the first time.

Case Report

This 25-year-old woman presented for evaluation with a 5-year history of chronic right-sided neck twitching. This symptom gradually diminished and then became prominent in her right shoulder, accompanied by weakness. Her medical history was noncontributory. She had no complaints of swallowing difficulties or voice problems at the time of her neurological evaluation. On physical examination, she was noted to have a right winged scapula, atrophy of the right sternocleidomastoid muscle, and weakness of right shoulder shrug. Electromyography showed absence of electrical activity in the sternocleidomastoid muscle. Imaging revealed a bone density dorsal to the hypoglossal tubercle (Figs. 1 and 2). Vascular disease was ruled out with formal angiography. A right lateral suboccipital craniotomy was performed while using evoked potential monitoring. The intraoperative finding was a small nonossous mass of densely calcified rootlets, which clearly appeared intradural. Provocative stimulation of the efferent rootlets did not elicit a response, and the lesion was amputated for pathological examination and processing. Histopathology showed inflammatory changes in the peripheral nerve with extensive calcification, predominantly amorphous type, without psammoma bodies or meningothelial whorls (Fig. 3). Immunohistochemical staining for S100 protein confirmed the presence of peripheral nerve.

Discussion

Previous case descriptions have only been of neuritis ossificans in peripheral systemic nerves. The origin or pathogenesis of this phenomenon is thought to be traumatic in nature. Neuritis ossificans is recognized as a process of heterotopic calcification confined to the epineu-
Neuritis ossificans of a cranial nerve

Repetitive injury and trauma seem to be major causative factors of neuritis ossificans in the peripheral nerve, and its pathogenesis appears to be similar to that of myositis ossificans. This mechanistic sequence consists of several phases: 1) trauma, 2) chronic inflammation, 3) fibroblast/osteoblast proliferation, and 4) ossification. However, chronic injury or irritation after mechanical trauma of a cranial nerve in this fashion is not a well-known phenomenon. The etiologies of isolated accessory neuropathy in the preimaging era were considered obscure in every report, but in the present case, a bone density was detected preoperatively in the posterior fossa ipsilateral to the deficit. The presence of cranial nerve rootlet ossification suggests that neuritis ossificans was caused by a repetitive mechanical trauma or friction of the nerve near the porous hypoglossus. Additionally, pathological analysis confirmed a zonal pattern of calcification characteristic of neuritis ossificans in cranial nerve rootlets accompanied by inflammatory changes, which represents a clue to the development of this uncommon pathology.

To date, only cases of neuritis ossificans involving peripheral nerves have been reported in the literature. This case study represents the first report of neuritis ossificans in a cranial nerve. The presentation of this lesion in the rootlets of a cranial nerve indicates no cytoarchitectural preference for this etiology between cranial and systemic nerves.

Conclusions

Neuritis ossificans is a very rare pathological entity that can affect both systemic peripheral and cranial nerves. Trauma, including tension of the nerve, appears to be the most significant causative factor in the development of this pathologic entity. The zonal calcification and inflammation of neuritis ossificans is similar to that seen in myositis ossificans and generally results in the destruction of the nerve, both anatomically and functionally.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.
Author contributions to the study and manuscript preparation include the following. Conception and design: Kemper. Acquisition of data: Kemper, Bauserman. Drafting the article: Kemper. Critically revising the article: Rojas, Bauserman. Reviewed final version of the manuscript and approved it for submission: Kemper.

References


Accepted May 3, 2010.
Please include this information when citing this paper: published online May 28, 2010; DOI: 10.3171/2010.5.091931.
Address correspondence to: Craig M. Kemper, M.D., 801 35th Street, Suite 400, Austin, Texas 78705. email: cmkmd@austin.rr.com.