Demyelinating Hypertrophic Inferior Alveolar Nerve Mimicking a Nerve Tumor

Hiroaki Fujita, Norito Kokubun, Tsubasa Sada, Takahide Nagashima, Tomoko Komagamine, Kiyokazu Kawabe and Koichi Hirata

Abstract

We herein report a patient with demyelinating inferior alveolar nerve hypertrophy, which was initially suspected to have a nerve tumor. A 39-year-old woman with childhood-onset polyneuropathy presented with tooth pain and visited a dental clinic. An X-ray examination of the mandible revealed enlargement of the mandibular canal, and a nerve tumor was suspected. CT scan and MRI showed hypertrophy of the inferior alveolar nerve along its entire length. We diagnosed the patient with chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), which was supported by the spontaneous recovery reported in her childhood, the results from a nerve conduction study and MRI data. CIDP should be considered in the differential diagnosis of mandibular canal enlargement.

Key words: chronic inflammatory demyelinating polyradiculoneuropathy, nerve tumor, mandibular canal, nerve hypertrophy, inferior alveolar nerve

(Intern Med 54: 1109-1111, 2015)
(DOI: 10.2169/internalmedicine.54.3512)

Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), an acquired immune-mediated peripheral nerve disorder, causes nerve hypertrophy. Approximately 70% of patients with CIDP have enlarged nerves in some part of the body (1). Spinal nerve roots and brachial and lumbar plexus are the most common sites of nerve hypertrophy (2). However, cranial nerve involvement in CIDP is rare. We herein report a patient with suspected CIDP and inferior alveolar nerve hypertrophy, which mimicked an intraosseous inferior alveolar nerve tumor.

Case Report

The patient was a 39-year-old woman who had presented with walking difficulty at age 3. She had no family history of neuromuscular disease and her physical growth and development from birth to 3 years of age were normal. Although she could not stand up and walk at age 4, her symptoms spontaneously recovered and she was to be able to walk again by age 6 with residual equinus feet and muscular atrophy in the distal extremities. She underwent an operation at age 12 to elongate the Achilles tendon. When she was 24 years old, she felt a tingling sensation in her left ring and little fingers. Nerve enlargement was detected in the upper arm segment of the ulnar nerve on the left side. A peripheral nerve schwannoma was suspected and an open biopsy was performed. However, the details of the biopsy were not available.

The patient visited our hospital at 33 years of age. An examination showed severe weakness and atrophy in her small hand muscles and lower legs. However, the proximal muscles were preserved. She did not present with pes cavus and walked with a steppage gait. The patient’s tendon reflexes were diminished in the legs but normoactive in the arms. Palpable, enlarged nerves, which corresponded to the ulnar nerve, were observed in the upper arms bilaterally. The patient had decreased light touch and pain sensations in her fingertips and distal part of the legs. Her cranial nerve functions and autonomic nervous system were intact. A nerve conduction study showed excessive temporal dispersions of proximal compound muscle action potentials in the median nerve.

1Department of Neurology, Dokkyo Medical University, Japan and 2Department of Neurology, Toho University Omori Medical Center, Japan
Received for publication June 19, 2014; Accepted for publication September 19, 2014
Correspondence to Dr. Norito Kokubun, kokubun@dokkyomed.ac.jp
Figure 1. Motor nerve conduction study in the median and ulnar nerves. Excessive temporal dispersions of proximal compound muscle action potentials (CMAPs) were recorded in the forearm segments of the median and ulnar nerves. Motor conduction velocities (MCVs) were also reduced (Normal, >50 m/s in the median and >52 m/s in the ulnar nerves). In contrast, distal motor latencies (DMLs) were nearly normal (normal: <4.1ms in the median nerve and <3.6ms in the ulnar nerve).

Figure 2. Radiographic features in the patient. A: Enlarged mandibular canals (arrows) on X-ray examination. B and C: CT scan showed an enlarged mandibular canal (arrows). Axial view (B) and coronal view (C). D: MRI of the mandible, axial image. Hypertrophic inferior alveolar nerves showed high signal intensity on a T2 weighted image (arrows). B: Reconstructed 3D-MRI. Massive, hypertrophic inferior alveolar nerves were detected, predominantly on the left. A 3D image of the inferior alveolar nerves was reconstructed with constructive interference in steady state (CISS) images and highlighted using the graphic processing software “AZE VirtualPlace” (AZE, Tokyo, Japan). Background structure was generated with magnetization prepared rapid acquisition with gradient echo (MPRAGE) images. A: anterior, R: right, L: left.
Peripheral nerve hypertrophy is one of the supportive diagnostic features of CIDP (2). However, other demyelinating neuropathies, such as CMT1, also cause nerve enlargement (3). The present case did not have a family history or a duplicated PMP22 gene or MPZ gene mutation. Moreover, spontaneous recovery in childhood and abnormal temporal dispersions in the intermediate nerve segment, which were recognized as “non-uniform demyelination”, are unlikely in hereditary demyelinating neuropathies. Cranial nerve hypertrophy is a rare manifestation of CIDP; involvement of the oculomotor (4) and trigeminal (frontal and infraorbital) nerves (4-8) has been previously reported. A pathology study found interstitial amorphous substances in the endoneurium and onion bulb formations in the enlarged nerves (9).

In the present case, hypertrophy of the intraosseous inferior alveolar nerves was revealed following a finding of an enlarged mandibular canal. An enlarged mandibular canal is commonly observed with several types of neoplasms (10, 11). Ethunandan et al. described a patient with a hypertrophic inferior alveolar nerve (12). An open biopsy was performed on this patient and onion-bulb formation and diffuse lymphocyte infiltration, particularly around blood vessels, were found (12). The patient was diagnosed with “localized hypertrophic neuropathy”. Although the patient presented with no neurologic deficits other than numb-chin syndrome, radiological information for other parts of the patient’s body was not available. In the present case, childhood onset was the possible cause of the bone change. Brachial plexus and ulnar nerve hypertrophy on MRI and ultrasonography supported the diagnosis of CIDP. CIDP should be considered as one of the causes of inferior alveolar nerve hypertrophy and mandibular canal enlargement.

The authors state that they have no Conflict of Interest (COI).

References