Post-Irradiation Myokymia and Neuromyotonia in Unilateral Tongue and Mentalis Muscles: Report of a Case

Lu-Han Liu, Chung-Wen Chen, and Ming-Hong Chang

Abstract- We report a 52-year-old man with slowly progressive dysarthria and dysphagia for about 11 years after radiation therapy of nasopharyngeal carcinoma. Neurological examination revealed atrophy and myokymia on the left side of the tongue and in the left mentalis muscles. Electrical discharges of myokymia and neuromyotonia were also observed in the aforementioned muscles, suggesting increased motor axonal membrane excitability involving the left hypoglossal nerve and the marginal mendibular branch of the left facial nerve. Magnetic resonance imaging of the brain did not show any evidence of tumor recurrence, indicating that irradiation probably plays an important role in pathogenesis. Focal myokymia with concomitant neuromyotonia in unilateral tongue and mentalis muscles could be an unusual delayed manifestation after radiation therapy.

Key Words: Myokymia, Neuromyotonia, Tongue, Mentalis muscle, Irradiation

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INTRODUCTION

Myokymia and neuromyotonia are clinical syndromes characterized by hyperexcitability of peripheral motor axons. Both may occur in a generalized or focal fashion and reflect a generalized or focal alteration in the peripheral nerve membrane itself or its microenvironment[1]. Focal myokymia could be associated with intrinsic brainstem lesions (mainly tumors and multiple sclerosis) and Guillain-Barré syndrome[2,3]. Radiotherapy-related cranial nerve palsy (most frequently in the hypoglossal nerve) may occur in patients with nasopharyngeal carcinoma (NPC) after conventional radiotherapy[4].

We describe an unusual patient with isolated, clinically and electrophysiologically manifest, myokymia and neuromyotonia on the left side of the tongue and in the left mentalis muscles after radiotherapy to the neck for NPC.

CASE REPORT

The patient is a 52-year-old man who was diagnosed with nasopharyngeal carcinoma (poor differentiated

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type) with bilateral lymph nodes metastasis (mainly on the left side) in late 1993. He was treated with Cobalt-60 radiotherapy (72 Gy TD/60 fractions/6 weeks) to the nasopharynx with bilateral parallel opposed portals and shrinking field. The NPC has been in remission ever since.

About 11 years later, he gradually developed difficulties in swallowing and limited motion of tongue. Five months after the onset of his symptoms, physical examination revealed dysarthria and dysphagia, as well as atrophy of the left half of the tongue and left mentalis muscles with continuous undulation (Figs. 1-3). There was also spontaneous contraction of the left mentalis but not the facial, masseter, or limb muscles. Thickened and hardened skin was noticed on both sides of the neck pre-

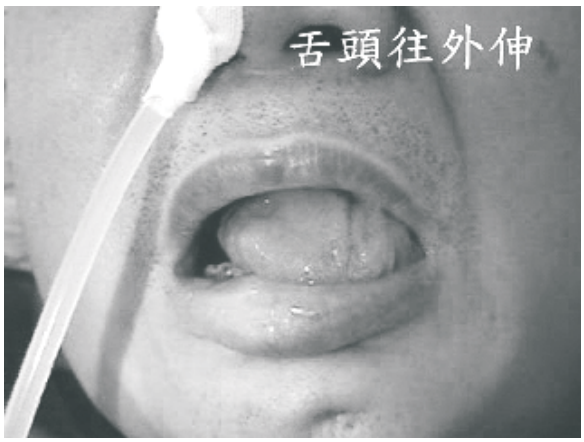

Figure 2. Atrophy and rippling of left mentalis muscle and left-sided tongue, and tongue deviation to left side on protrusion.

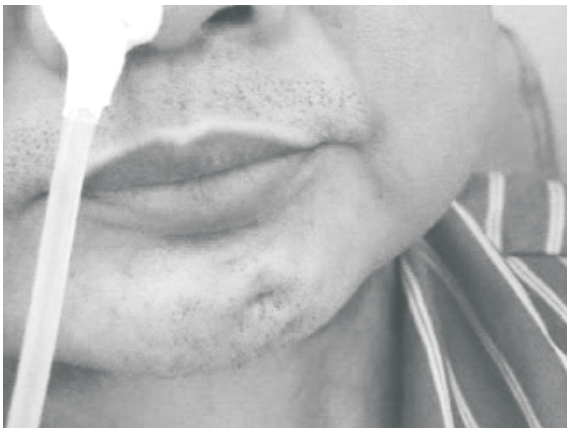

Figure 1. Atrophy and indentation of left mentalis muscle.

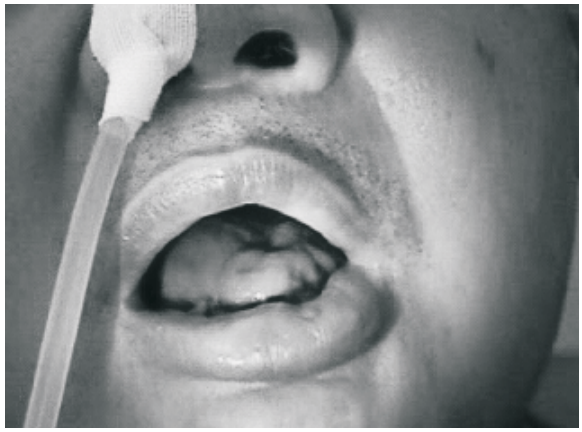

Figure 3. Atrophy and undulation of left-sided tongue when opening mouth.

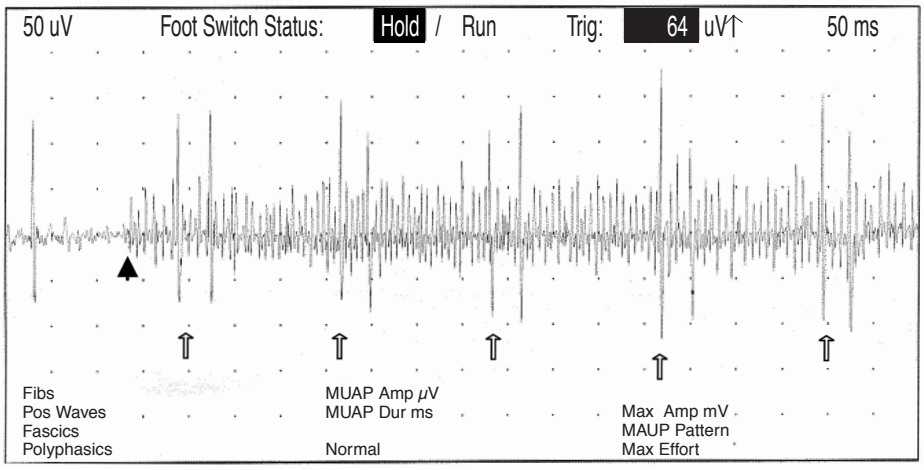

Figure 4. Myokymic and neuromyotonic discharges recorded from left mentalis muscle. The black arrow head was start of neuromyotonic discharges. The white arrow head indicates continuous type myokymic paired discharges.

sumably due to previous radiotherapy (more on the left side). There is also right hemiparesis (MP: Gr.5-/5) which is ascribable to an old cerebral infarction 3 years ago. The rest of the examination was unremarkable.

Needle electromyogram (EMG) of the tongue and left mentalis muscles at rest showed two patterns of motor units firing, one with a firing frequency of around 5 Hz, consistent with myokymia (continuous type), and the other with a firing frequency of around 150 Hz, consistent with neuromyotonia (Fig. 4). Results of magnetic resonance imaging of the head showed normal contour of the nasopharynx without any contrast-enhanced mucosal mass lesion. The adjacent bone marrow signal is well preserved without tumor infiltration, so is the skull base or the cranial fossa base under the Meckel's cave. Also, biochemical tests showed no significant abnormal findings.

DISCUSSION

No more than 40 patients with NPC who had cranial nerve palsy subsequent to conventional radiation therapy have been reported in the English literature since 1966[4]. There are only rare reports of myokymia and neuromyotonia in these patients. Wang et al[5] described tongue myokymia in a patient who had radiation therapy for NPC. The patient reported here had focal myokymia and neuromyotonia involving both facial and hypoglossal nerves as a late complication of irradiation. The distribution is previously unreported.

Our patient had atrophy and myokymia/neuromyotonia in the left half of the tongue and left mentalis muscles. Tongue and mentalis muscle atrophy implicate the involvement of the intramedullary or extramedullary portion of the hypoglossal nerve and the marginal mandibular branch of the facial nerve, respectively. It suggests that the nerves located close to the left side of the tongue and the left submandibular region are affected. Since MRI of head in our case did not show any evidence of recurrence of tumor, the nerve injury was most likely ascribable to a delayed effect of irradiation. The dose of Cobalt-60 radiotherapy is 70 Gy in our patient, and a dose range from 62.5 to 100 Gy has been reported to induce cranial nerve lesion[6]. If it is the case, then the latency between irradiation and cranial nerve palsy is 11 years in our patient (12 to 172 months in previous reports).

Radiation-induced delayed nerve damage is thought to result from a direct damage to the nerve cells, or alternatively, from a damage to the vascular endothelium and consequent ischemia of the adjacent tissues. Connective tissue fibrosis and secondary nerve injuries may then ensue[6]. The degree of damages appears to be positively, whereas the time of onset of the neurologic signs appears to be inversely, correlated with the radiation dose[6]. Both myokymia and neuromyotonia are thought to originate in segments of the motor axon, and are presumed to result from spontaneous depolarization or ephaptic transmission secondary to the altered membrane excitability in the demyelinated nerve[2,7].

Alternations of peripheral nerve voltage gated K+ channels (VGKC) have recently been associated with several examples of both myokymia and neuromyotonia. For example, VGKC antibodies were found in some cases of generalized neuromyotonia (Isaacs' syndrome)[8] and persistent facial myokymia[9]. Whether abnormalities of VGKC also serve as a mechanism underlying the other disorders with myokymia and neuromyotonia remains to be determined[10].

Myokymia and neuromyotonia have different clinical and electrophysiologic manifestations[2,11], but probably share a common mechanism of excessive motor unit activities presumably originating from the motor axon[11]. Myokymia refers to the clinical phenomena of undulating or rippling muscles, as if "tiny snakes wriggle beneath the skin". Because myokymia is a disorder of the motor unit, it may occur transiently for days to months, e.g. facial myokymia of multiple sclerosis and Guillain-Barré syndrome, or may persist for many years, e.g. pontine glioma and post-irradiation. On the other hand, neuromyotonia is usually characterized by insidious-onset but persistent contraction of the affecting muscles. Clinical myokymia and neuromyotonia are evident in our patient.

Two EMG patterns, myokymic and neuromyotonic discharges (Fig. 4), were documented in our patient.

Myokymic discharges may be present continuously or discontinuously[12]. There are characteristic rhythmic discharges of a single motor unit potential or semi-rhythmic stereotyped bursts of motor unit potentials. The frequencies within the burst are 20-40 Hz, and the intervals between bursts are 0.25 to 4 seconds. In the continuous type, there are rhythmic single or paired discharges of one or a few motor units recurring at a fairly uniform rate of 1-5 Hz. In the discontinuous type, there are bursts of motor unit activities at 20-40 Hz, lasting for 100-1000 ms and alternating with pauses up to a few seconds[12]. Neuromyotonic discharges are muscle action potentials from one or several muscle fibers[13], and usually are associated with myokymic discharge[12]. They fire in bursts with frequencies higher than 100 Hz for 0.5-2.0 seconds[12], and the bursts often start and stop abruptly[13]. Because of fatigue of individual muscle fibers, the amplitude may gradually decrease (i.e. the waning form)[13].

Our report revealed that focal myokymia and neuromyotonia in unilateral tongue and mentalis muscles could be an unusual delayed complication of radiation therapy.

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