Cheirobuccopedal Syndrome

Wei-Hsi Chen, Yi-Shan Wu, and Hung-Sheng Lin

Abstract-

Objectives: Restricted sensory syndrome provides an excellent chance for the understanding of neuroanatomic correlation. The trigeminal nerve has been shown to convey buccal sensory impulse to central compartment. However, the pathway between cortex and the trigeminal sensory nucleus remains largely unknown.

Method: A patient presented with cheirobuccopedal syndrome, or objective sensory impairment confined to the left intraoral cheek, hand and foot, was reported.

Results: Decrease in pinprick pain and fine touch sensation was detected at the left intraoral cheek, hand and foot. A recent infarction was disclosed at the left paramedian pons. The foregoing hypalgesia and hypesthesia recovered within one month after onset.

Discussion: The trigeminobuccal sensory tracts are deemed to run in parallel with other spinothalamic and trigeminothalamic tracts within the brainstem. The rarity of buccal sensory deficit upon brainstem damage may be due to dispersion of the trigeminobuccal sensory tracts on their way of ascending, or their relatively high tolerance to different insults.

Key Words: Intraoral cheek, Sensory, Pons, Trigeminobuccal tract, Trigeminothalamic tract, Trigeminal sensory nucleus, Lateral spinothalamic tract, Cerebral infacrtion

Acta Neurol Taiwan 2009;18:34-36

INTRODUCTION

Different types of restricted sensory impairments or incomplete forms of pure sensory stroke have been reported after the advent of magnetic resonance technology. providing novel insight into neuroanatomic pathways and clinical outcome. Cheiro-oral syndrome (COS)[1] and cheiro-oral-pedal syndrome (COPS)[2] are characterized by sensory impairment confined to ipsilateral mouth angle, finger(s)/hand, and/or foot. These sensory syndromes call for attention due to involvement of two or more body topographies in disparity, a probable disproportion between size of pathology and sensory deficit, and most importantly, an emerging presentation

From the Department of Neurology, Chang Gung Memorial Hospital-Kaohsiung Medical Center, and College of Medicine, Chang Gung University, Kaohsiung, Taiwan.
Received June 27, 2008. Revised July 7, 2008.
Accepted September 1, 2008.

Reprint requests and correspondence to: Hung-Sheng Lin, MD. Department of Neurology, Chang Gung Memorial Hospital, No. 123, Tai Pei Road, Niao Sung Hsiang, Kaohsiung 833, Taiwan.
E-mail: a2522kimo@yahoo.com.tw

of neurological disorder since COS was firstly mentioned at 1914[3]. A variety of related variants of COS and COPS have been reported, such as cheiroretroauricular syndrome, cheirobuccal sensory syndrome[4] and crossed cheiro-oral syndrome[5]. We report a patient who presented sensory impairment at ipsilateral intraoral cheek, hand and foot distinct from the related cases described before.

CASE REPORT

A 48-year-old man experienced an abrupt onset of numbness confined to his left hand, foot and intraoral cheek in one late morning. He had had hypertension for two years without treatment, and caught common cold one week before. Otherwise, he denied craniofacial trauma, migraine or other disorder.

He consulted our department on the next day due to persistent numbness. His blood pressure was 150/85 mmHg. He was conscious and oriented. Neurological examination revealed a subjective 80% decrease of pinprick pain and 50% decrease of cotton fine touch at left intraoral cheek, hand and foot; and 50% decrease of vibratory sense at left hand and foot. There was no sensory deficit detected at mouth angle or face. Corneal reflex and positional joint sensation were preserved. Two-point discrimination test (reference range: 8-15 mm; average 10 mm) showed a mild impairment in the left hand (left palm 12 mm; right palm 10 mm) and foot (left foot 14 mm; right foot 10 mm). Baroesthesia and stereoesthesia were not detected. Magnetic resonance image showed a high diffusion-weighted (Fig. A) and T2-weighted intensity (Fig. B) at left paramedian pons, compatible with a recent infarction. The size of infarction was 159.6 mm^3, which was larger than lacunae[6]. Magnetic resonance angiogram disclosed patent major intracranial arteries. Stroke risk factor survey revealed low blood high-density-lipoprotein- cholesterol (36 mg/dl), systolic hypertension, left ventricular hypertrophy, ST-T elevation, and obesity (body mass index = 35.1). His sensory deficits in the left intraoral cheek, hand, and foot subsided within one month. He did not experience any more stroke events or sensory deficits afterwards.

DISCUSSION

Although intraoral cheek contains abundant sensory receptors, buccal sensory impairment is a rare neurological symptoms found in central lesions. Generally, sensory impairment of the intraoral cheek occurs with lesions in the unilateral trigeminal nerve[7] or sensory nucleus[8-9], and rarely, contralateral pons[10] or ventral thalamus[4]. In these cases, patients usually suffer additional sensory impairment in the face or body, indicating co-localization of sensory tracts from intraoral cheek and other dermatomes. As the findings of this patient also indicates, the centripetal trigeminobuccal sensory tracts enter into brainstem by means of trigeminal nerve and nucleus. The tracts then cross to the other side and ascend in

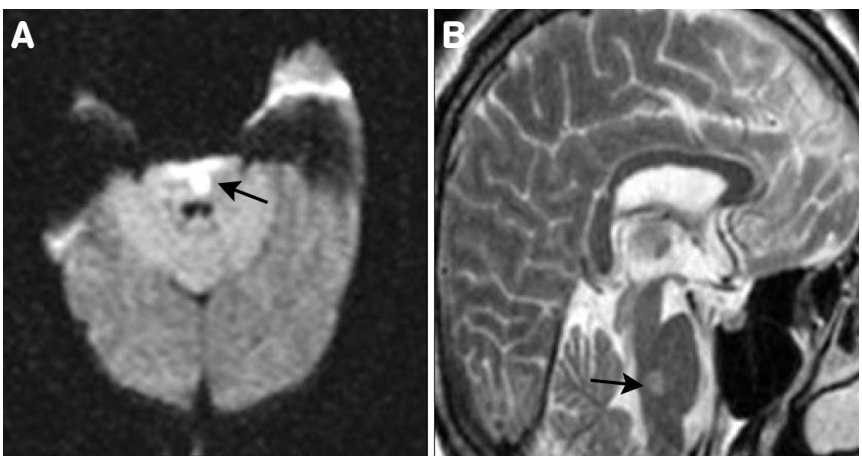

Figure. Magnetic resonance image showed a high diffusion-weighted (A) and T2-weighted intensity, (B) at left paramedian pons, compatible with a recent infarction, (arrows).

company with the spinothalamic and trigeminothalamic tracts to reach the ventral thalamus. A compression of proprioceptive pathway by the edematous changes around the infarction may explain the impairment of vibratory sense in this patient and previously reported brainstem COS patients.

If the ascending trigeminobuccal sensory tracts run in parallel with spinothalamic and trigeminothalamic tracts, it may be hard to envisage why buccal sensory impairment present in this patient should be a rare occurrence. We would forward two explanations for this enigma. First, although the trigeminobuccal, trigeminothalamic and spinothalamic tracts run together within brainstem before arrival of thalamus, a dispersion of the trigeminobuccal sensory tract but not the trigeminothalamic and spinothalamic tracts may lead to a lower frequency of buccal sensory impairment. Second, the cooperation of neuronal vulnerability, sensory plasticity and preconditioned stress of the relevant areas at cortex[11] has been proposed to explain cortical COS. A similar condition may be present in pontine damage.

Our patient has a favorable outcome. Restricted sensory syndromes usually are associated with cerebral infarction and have favorable outcomes, probably because of a selective involvement of small penetrating arteries in the majority of patients. Concurrently, large cerebral infarction[11], large cerebral hemorrhage[11], medullary involvement[5], and basilar artery thrombosis[12] would predict exacerbation of neurological deficits and relatively poor outcomes in COS and other related restricted sensory syndrome. However, an accurate prediction of exacerbation in COS and related variants is still not possible because there is no ideal clinical marker suitable for exclusion. Neuroimage study thus should always be done for cases of COS and related variants although they usually result from lacunar stroke[13].

REFERENCES

1. Isono O, Hirayama K. Cheiro-oral syndrome [Jpn]. Nippon Rinsho 1993;51 Suppl:537-43.
2. Chen WH, Lin HS, Su CS, et al. Cheiro-oral-pedal syndrome [Chin]. J Kaohsiung County Med Assoc 2005;16:15-8.
3. Sitting O. Klinische Beitrage zur Lehre von der Lokalisation der sensiblen Rindenzentren. Prager Med Wochenschr 1914;45:548-50.
4. Wu YS, Li TH, Chen TH, et al. Cheirobuccal sensory syndrome. Acta Neurol Taiwan 2008;17:50-3.
5. Chen WH, Li TH, Chen TH, et al. Crossed cheiro-oral syndrome. Clin Neurol Neurosurg 2008;110:1008-11.
6. Bogousslavsky J, Orgogozo JM. Lacunar syndromes. In: Vinken PJ, Bruyn GW, Klawans HL, Toole J Handbook of Clinical Neurology-Vascular Diseases. North-Holland, Amsterdam, 1988;54(part II):235-70.
7. Penãrrocha M, Cervello MA, Marti E, et al. Trigeminal neuropathy. Oral Dis 2007;13:141-50.
8. Ferro JM. Intraoral sensory loss as the presenting feature in pontine hematoma. Cerebrovasc Dis 2000;10:486-7.
9. Graham SH, Sharp FR, Dillon W. Intraoral sensation in patients with brainstem lesions: role of the rostral spinal trigeminal nuclei in pons. Neurology 1988;38:1529-33.
10. Combarros O, Dóez C, Cano J, et al. Ataxic hemiparesis with cheiro-oral syndrome in capsular infarction. J Neurol Neurosurg Psychiatry 1992;55:859-60.
11. Chen WH, Lan MY, Chang YY, et al. Cortical cheiro-oral syndrome: a revisit of clinical significance and pathogenesis. Clin Neurol Neurosurg 2006;108:446-50.
12. Chen WH, Lan MY, Chang YY, et al. Bilateral cheiro-oral syndrome. Clin Neurol Neurosurg 1997;99:239-43.
13. Ivo L. CT scanning can differentiate between ischaemic attack and haemorrhage. BMJ 1999;319:1197-8.