Cheirobuccal Sensory Syndrome

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Abstract- Two patients presented with sensory impairments confined to their right intraoral cheek and right first three fingers. An objective decrease of pinprick pain was detected at these sites. Neuroimaging illustrated recent infarcts in the contralateral ventral thalamus of both patients. The intraoral and cheiral sensory impairments resolved within two months after onset. We coined the term "cheirobuccal sensory syndrome" to describe these cases. The clinical significance and pathogenesis of this peculiar syndrome are discussed.

Key Words: Thalamus, DWI, Infarct, Intraoral, Finger

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INTRODUCTION

Pure sensory stroke is the third most common lacunar syndrome. In contrast to a complete hemibody involvement in pure sensory stroke, incomplete sensory impairment confined to a restricted region or multiple body parts is occasionally reported, such as cheiro-oral syndrome[1], cheiro-oral-pedal syndrome[2], cheiroretro-auricular syndrome[3], thoracic segmental syndrome or pure trigeminal sensory syndrome[4]. Reports of these syndromes have been increasing, especially since the advent of magnetic resonance imaging. These particular sensory syndromes provide an opportunity to better understand the neuroanatomy and its sensory localization in clinical practice[5]. Herein, we report an unusual sensory disorder restricted to the intraoral mucosa and homolateral fingers due to a contralateral thalamic infarct. The pathogenesis of this peculiar syndrome is discussed.

CASE REPORT

Case 1. A 58-year-old man had a sudden onset of tingling and numbness sensations over his right intraoral cheek and thumb when he was talking. He consulted us the next day. He had been taking antihypertensive medications for four years but had discontinued them two months before this episode. He denied having a history of symptomatic stroke, coronary artery disease, recent head injury or illicit drug use. He consumed cigarette and alcohol socially for twenty years. On presentation, his blood pressure was 170/95 mmHg. He was obese, conscious and oriented. His eyegrounds showed grade I hypertensive arteriopathy. Abnormal neurological findings were a 30% hypesthesia to pinprick and fine touch over his right intraoral cheek and first three fingers. Vibratory sense, positional joint sensation and cortical sensory modality were normal. Taste, olfaction and

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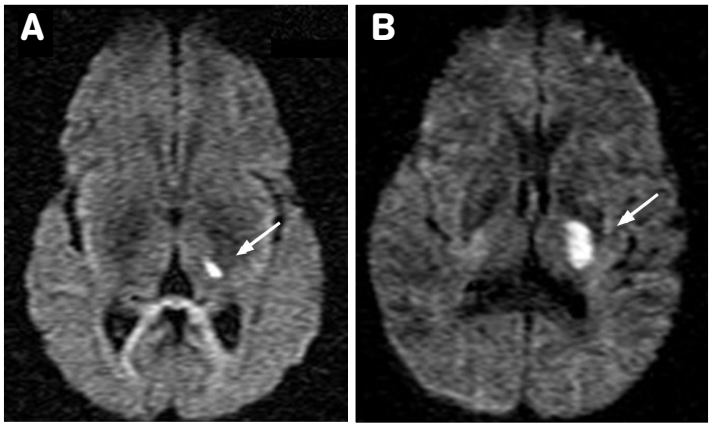

Figure 1. Magnetic resonance imaging of brain revealed a recent infarction (arrow) in the left ventral thalamus in Case 1 (A) and Case 2 (B).

hearing were not decreased. Ataxia was not detected. There was no carotid bruit. A stroke risk factor survey[6] revealed mild hypercholesterolemia (total cholesterol 220 mg/dl, low-density-lipoprotein- cholesterol 167 mg/dl). Head magnetic resonance imaging (MRI) revealed a high diffusion-weighted MRI (DWI) hyperintensity compatible with recent infarction at the left ventral thalamus (Fig. 1A), and diffuse narrowing of the intracranial arteries compatible with atherosclerosis. His intraoral and cheiral sensation recovered two weeks and eight weeks later, respectively. He remained well and has not experienced cardiovascular complications since that episode.

Case 2. A 62-year-old woman had a sudden onset of tingling and numb sensation over her right intraoral cheek and right hand first two fingers while watching television. She had received antihypertensive medication regularly for three years. She denied having a history of symptomatic stroke, coronary artery disease, recent head injury, illicit drug use, cigarette or alcohol consumption. On presentation, her blood pressure was 145/78 mmHg. She was conscious and oriented. Her eyegrounds showed grade I hypertensive arteriopathy. Abnormal neurological findings were 20% hypesthesia to pinprick pain and cotton fine touch over her right intraoral cheek and first two fingers of her right hand. Vibratory sense, positional joint sensation and cortical sensory modality were normal. Taste, olfaction and hearing were not decreased. Ataxia was not detected. There was no carotid bruit. A stroke risk factor survey[6] revealed a mild hypertriglyceridemia (serum triglycerides 186 mg/dl) and homocysteinemia (14.7 μg/ml). Head MRI disclosed a hyperintense DWI lesion compatible with recent infarction at the left ventral thalamus (Fig. 1B). Her intraoral and cheiral sensation recovered within one week and four weeks later, respectively. She remained well and has not experienced cardiovascular complications since that episode.

DISCUSSION

Restricted sensory impairment is not uncommon in central lesions[7]. However, sensory deficits confined to the intraoral cheek and homolateral distal hand has not been described previously. We coined the term "cheirobuccal sensory syndrome" to describe this condition. The corresponding infarctions were located in the lateral thalamus (ventroposterior medial nucleus and ventroposterior lateral nucleus of the thalamus) which is irrigated by a group of inferolateral arteries between the medial and lateral posterior choroidal arteries, arising from the posterior cerebral artery[8]. In addition to affecting sensory disturbances, occlusion of these arteries can also cause hemiataxia, hemiparesis and post-stroke central pain syndrome, depending on the involvement of the corresponding branch within this group of arteries[8].

There are a large number of sensory receptors in the mucosa of the intraoral cheek, and their impulses are conveyed through the trigeminal nerve and sensory nucleus similar to other craniofacial structures. However,

the precise ascending route of their second neurons is actually ambiguous when compared to our knowledge of the itinerary of the spinothalamocortical or trigeminothalamocortical tracts conveying sensory impulses from other structures. Sensory impairment of the intraoral cheek has only been reported in cases with involvement of the trigeminal nerve[9] or sensory nucleus [10-11] but not at other sites above them. This perplexity is exasperated when direct stimulation of the post-central or pre-central gyrus does not elicit a corresponding sensation in the intraoral cheek in humans, although experiments have shown a representative area of the intraoral cheek has been found near the oropharyngeal region in the sensory cortex[12]. The findings in our patients therefore support the notion that the second sensory neuron of the intraoral cheek reaches the ventral thalamus along with the ascending fibers from other cranial and somatic structures.

In our patients, a corresponding infarction was clearly identified in the contralateral ventral thalamus, indicating a crossed ascending sensory pathway between the intraoral cheek and thalamus. Although this neuroanatomic relationship has not been mentioned previously, transient dysesthesia of intraoral structures such as the tongue, throat, lips or cheek has occasionally been reported after stereotaxic thalamotomy in Parkinsonians[13]. Cambarros et al[14] have reported a patient having isolated sensory disturbance involving the left intraoral cheek, perioral region and tips of the left thumb and forefinger. A small infarct was found in the contralateral ventral thalamus. These clinical events adequately support an argument that there is an ascending sensory tract of the intraoral cheek to the contralateral ventral thalamus in humans.

Since the body topography arrays constantly within the ascending sensory tract or central nucleus, salutatory sensory impairment not only provides a localizing value but also calls attention to the pathogenesis of sensory expression among different neurons. In cheiro-oral syndrome or cheiro-oral-pedal syndrome, which presents with a restricted sensory involvement of the cheiral, oral or pedal region, a stereometric proximity of these sensory fibers in the thalamus[15], or a co-operation of neuronal

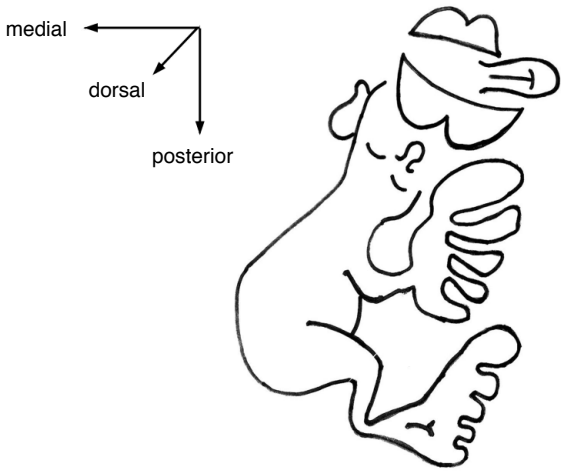

Figure 2. Stereometric diagram of the thalamus showing facial and bodily topography. The sensory neuron of the intraoral cheek is proposed to locate medially and dorsally.

vulnerability, sensory plasticity and preconditioned stress of these representative areas at cortex[16] have been proposed to explain these peculiar sensory syndromes. In our patients, restricted sensory impairment of the intraoral cheek and fingers implies two possibilities. First, proximity of these sensory fibers within the ventral thalamus is possible. If so, the location of the intraoral cheek sensory fibers should reside medially and dorsally in order to evade other acral sensory fibers (Fig. 2). Second, pre-existing damage of the intraoral cheek sensory fibers is superimposed on a new thalamic infarction which causes only a suboptimal threshold potential for other sensory neurons. Therefore, the sensory impairment is limited to the intraoral cheek and first two fingers. The hypertensive history of these two patients reinforces this possibility.

Nevertheless, intraoral cheek sensory impairment is rare in the clinical setting and is still somewhat a mystery. In addition to the above two proposals, we suggest a third possibility. The ascending sensory fibers of the intraoral cheek may disperse rather than converge. If this is true, it could explain why an involvement of the intraoral cheek is relatively infrequent in localized lesions. Indeed, modern techniques have already proven an inte-

gration of sensory fibers from neighboring structures terminating at the cortical representative area of a specific topography. This possibility should be challenged by more proofs.

Cheiro-oral or cheiro-oro-pedal syndromes usually have favorable outcomes. A deterioration of the clinical condition is infrequent except with a large hemorrhage, infarction or tumor[17-18]. In our two patients, sensory impairments resolved completely within three months after onset. It is obvious that the prognosis of these restricted syndromes depends on the underlying cause rather than the neurological presentation itself.

Our findings demonstrate that the localization of what we called the cheirobuccal sensory syndrome is at the contralateral ventral thalamus, and they also depict the connection between the intraoral cheek and thalamus. However, conclusions about clinical significance and prognosis should be based on supportive future findings/cases.

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