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Crossed cheiro-oral syndrome

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ABSTRACT

Objectives: Cheiro-oral syndrome is characterized by sensory impairment confined to perioral area and ipsilateral fingers/hand. It results from an involvement of the ascending sensory tracts above the pons. However, a crossed pattern of perioral and acral paresthesia was rarely reported before.
Patients and methods: This study reports the neuroanatomic relationship, course and clinical significance of perioral and contralateral acral paresthesia in four patients. We term it the crossed cheiro-oral syndrome.
Results: All patients had lateral or dorsolateral medullary infarctions that were ipsilateral to their perioral paresthesia. The contributory origin is considered a diagonal lesion involving the par oralis fibers within the descending trigeminal sensory tract and acral portion of the lateral spinothalamic tract at the lateral portion of medulla oblongata. Despite of a restricted sensory disturbance at initial, progressive neurological disability terminated to Wallenberg's syndrome ensued in three patients and disabling deficits persisted in two of them.
Conclusion: The crossed cheiro-oral syndrome seems a mild form of Wallenberg's syndrome. Therefore, it predicts medullary involvement and is also a warning sign for progression.

1. Introduction

Cheiro-oral syndrome (COS), first reported in 1914 by Sitting [1], is a clinical entity characterized by sensory impairment confined to the perioral area and ipsilateral finger/hand. Initially, this peculiar sensory syndrome was believed to have a localizing capacity which gradually burns down when an involvement of the ascending sensory tracts at the cortex [2], corona radiata, midbrain, thalamus or pons [3] can cause unilateral COS. Only the bilateral COS is still considered predictive of detriment at paramedian pons [4] and in rare case, the thalamus [5]. Nevertheless, COS offers a noteworthy substantial of understanding the clinical significance in practice, anatomic arrangement of sensory tracts and plasticity of sensory neurons [2,5]. We encounter four patients whose sensory impairments were restricted to the perioral area and contralateral distal hand. This crossed sensory impairment has not been mentioned before. We term it the crossed COS. The corresponding lesions were exclusively at the medulla oblongata in them. Neurological function subsequently deteriorated in three patients. The neuroanatomic relationship, course and clinical significance of this crossed COS are discussed.

2. Patients and methods

This study reports four patients with initial neurological defects of sensory impairment at their perioral area and contralateral fingers/hand. These four patients were not enrolled in our database of COS, which included patients with sensory impairment confined to their perioral area and ipsilateral distal fingers/hand without detectable defect in mentality, memory, motor or cerebellar function examined by neurologist [2,5]. We refer to this particular presentation investigated in this study as crossed COS. All patients denied migraine, craniofacial trauma within the previous 3 months, recent infection or illicit drug use. Stroke risk factor survey, lupus study, echocardiogram, and neurodiagnostic studies, including cranial computerized tomography or head magnetic resonance imaging with magnetic resonance angiography, somatosensory evoked potential, carotid duplex and transcranial doppler, were performed in all patients as described in our previous works [2,5]. Cerebral angiography was optional. The neurological status was assessed by the modified ranking scale (MRS).

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3. Results

3.1. Case 1

A 53-year-old man was admitted the day after experiencing a sudden onset of rotatory vertigo, facial flushing, throbbing headache and tingling sensation at his left mouth corner and thumb and forefinger of right hand on the morning while he was working in office. Hypertension has been diagnosed 5 years earlier. Neurological examination revealed a decrease of sensitivity to pinprick pain at his left mouth corner and right first three fingers. Cranial computerized tomography revealed no consistent abnormality. Besides of a deterioration of tingling sensation at mouth corner and fingers, vomiting, left Horner's syndrome, ataxic gait, and paresthesia of left arm and bilateral faces developed within 6 h. Head magnetic resonance imaging showed a high DWI intensity, which was compatible with an infarction, at the left dorsolateral medulla oblongata (Fig. 1A). Magnetic resonance angiography revealed mild atherosclerosis of posterior circulation. Stroke risk factor survey additionally disclosed hypercholesterolemia (265 mg/dl). His sensory impairment subsided within 3 months but his daily activity was limited by disabling ataxia. The MRS was 3 scores 5 years later.

3.2. Case 2

A 41-year-old woman experienced a sudden onset of floating sensation, nausea, palpitation, unsteadiness and tingling sensation at her left mouth corner, upper and lower lip, and the first three fingers of right hand 1 h after awakening from sleep on morning. She was treated at emergent care 20 min later. Hypertension, diabetes mellitus and dyslipidemia had been diagnosed 3 years earlier. Neurological examination revealed a decrease of sensitivity to pinprick pain at left mouth corner, upper and lower lip, and all fingers on the right hand. Cranial computerized tomography showed no consistent abnormality. Besides of a deterioration of her tingling sensation at mouth corner and fingers, rotatory vertigo, vomiting, left Horner's syndrome, left hyperhydrosis, choking, tongue weakness, and paresthesia over bilateral faces and right hemibody rapidly developed within 3 h. Head magnetic resonance imaging disclosed a high T2-weighted intensity, which was compatible with an infarction, at the left dorsolateral medulla oblongata (Fig. 1B). Magnetic resonance angiography revealed mild atherosclerosis in both carotid and vertebrobasilar arterial system. Stroke risk factor survey further disclosed homocysteinemia (17.0 U/ml) and protein S deficiency (45%). She recovered from residual vertigo and left perioral paresthesia, which responded well to amitriptyline but not clonidine. During the following 3 years, she experienced a similar episode. Her MRS was 1 score. Repeated magnetic resonance imaging revealed no new lesion.

3.3. Case 3

A 50-year-old man was admitted the day after experiencing a sudden onset of floating sensation and numb sensation at the right mouth corner and the left thumb on the morning while he was talking. Although hypertension had been diagnosed for 2 years earlier, he did not seek treatment. Neurological examination revealed a decrease of sensitivity to pinprick pain at his right mouth corner and left thumb. Head magnetic resonance imaging showed a high T2-weighted intensity, which was compatible with an infarction, at the right lateral medulla oblongata (Fig. 1C). Magnetic resonance angiography revealed mild atherosclerosis of posterior circulation. Stroke risk factor survey further disclosed homocysteinemia (15.0 U/ml). His sensory disturbance subsided 2 months later. His MRS was 1 score 5 years later.

3.4. Case 4

A 63-year-old man developed dizziness, fatigue and piercing pain at the right mouth angle and the left thumb at afternoon while he was doing farm work. He was admitted with these symptoms 2 days later. Hypertension was known for 10 years. Neurological examination revealed a decrease sensitivity to pinprick pain at the right mouth angle and first two fingers of left hand. Cranial computerized tomography showed no consistent abnormality. He refused further treatment and insisted on discharge despite right Horner's syndrome was detected 2 h after cranial computerized tomography. However, rotatory vertigo, vomiting and hyperhidrosis developed the next day. Head magnetic resonance imaging disclosed a high DWI intensity, which was compatible with an infarction, at the right dorsolateral medulla oblongata (Fig. 1D). Magnetic resonance angiography showed a diffuse intracranial and precerebral vascular stenosis. Stroke risk factor survey further disclosed homocysteinemia (15.5 U/ml) and hypercholesterolemia (235 mg/dl). After 1 year, he recovered from dizziness and right perioral dysesthesia, which had responded well to amitriptyline but not clonidine, and returned to farm work with residual ataxic gait. His MRS was 1 score 4 years later.

4. Discussion

The classical COS is an incomplete form of pure sensory stroke. It arises from an involvement of the spinothalamocortical and trigeminothalamocortical sensory pathway containing afferents of pars oralis and pars cheiralis between the cortex and pons, results from lacunar infarction in most situations, and usually terminates with favorable outcome [6]. On contrary, although crossed COS is also an incomplete pure sensory stroke, it strongly predicts an involvement of mediolateral medulla oblongata, associates with a sizable infarction, and usually carries an unfavorable prognosis. These differences notify a different clinical significance, vascular pathogenesis, and outcome in crossed COS from classical COS.

In the medulla oblongata, a dense and orderly array of sensory fibers (from lateral and anterior spinothalamic tract, medial lemniscus, and ventral and dorsal trigeminothalamic tract) and nuclei (spinal trigeminal nucleus and medial lemniscus) establish the sensory network governing the bodily somatic sensation. Importantly, the decussation of these sensory fibers offers the advantage of a crossed pattern of sensory disturbance in medullary involvement.

Six common sensory impairment patterns reportedly attributable to involvement of the medulla oblongata are ipsilateral trigeminal–contralateral limb/body, contralateral trigeminal–contralateral limb/body, bilateral trigeminal–contralateral limb/body, isolated limb/body, isolated ipsilateral trigeminal, and isolated contralateral trigeminal. They result from an involvement of the lateral medulla oblongata [7–9]. Other different combinations of sensory deficits at face, limbs and trunk, unilaterally or bilaterally, have been described [7,10–12]. A restricted sensory impairment confined to segmental [13] or dermatomal distribution [14] at limb/body has also been mentioned. However, a restricted sensory impairment as in our patients has not been reported before. The sensory distribution in our patients resemble a variant of the ipsilateral trigeminal–contralateral body/limb pattern or Stopford type I sensory loss which indicates an involvement of lateral part of medulla oblongata [15], but differs in a restriction of sensory distribution and an absence of motor weakness. Accordingly, crossed COS is a variant and mild form of Wallenberg syndrome.

Reviewing the literature, there are three patients to have a crossed sensory impairment confined to the perioral area and con-