

Opercular Cheiro-oral Syndrome

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● Perioral and distal upper limb sensory dysfunction (cheiro-oral syndrome) has classically been attributed to cortical involvement. In previously reported cases of the syndrome, caused by stroke, however, the thalamus or brain stem has been the actual site of the lesion. We have studied two patients with infarct in the superficial middle cerebral artery territory involving the parietal operculum. Sensory involvement was purely subjective in the face, but severe hypoesthesia was present in the distal upper limb, involving mainly position sense, stereognosis, and graphesthesia. Temperature and pain sensation were involved in one patient. These findings correlated with involvement of the lower part of the postcentral gyrus, more caudal parts of the parietal operculum, and underlying white matter. This opercular cheiro-oral syndrome seems more uncommon than faciobrachiorural hemihypoesthesia associated with anterior parietal artery territory infarct. A double supply to the parietal opercular region through branches of the temporal arteries and anterior parietal artery may explain the rarity of cheiro-oral syndrome resulting from hemisphere stroke, because simultaneous and partial compromise to two different pial artery networks is uncommon.

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The "operculum of the insula of Reil," or operculum, refers to the region that surrounds the insula (*operculum* means lid or cover). Since it is formed by part of the frontal, temporal, and parietal gyri, the operculum is usually divided into a frontal operculum,

which includes the lower part of the precentral gyrus and of the posterior part of F_3 , a temporal operculum, which is the rostral part of the superior temporal gyrus up to Heschl's transverse gyrus, and a parietal operculum, which includes the lower part of the postcentral gyrus and P_1 , and may extend to the rostral part of P_2 . While an anterior operculum syndrome due to involvement of the frontal operculum has been recognized,^{1,4} the neurologic picture associated with damage to the posterior part of the operculum has not been adequately studied. We describe two patients with sensory involvement limited to the perioral region and distal part of the upper limb (cheiro-oral syndrome), in association with parietal opercular infarction.

REPORT OF CASES

CASE 1.—A 63-year-old right-handed male engineer was a cigarette smoker and had untreated hypercholesterolemia. One morning at 2 AM, he suddenly experienced a feeling of disappearance of his right forearm, associated with a decrease of touch sensation without any weakness. At the same time, he experienced tingling in the right side of his mouth. He was admitted to hospital 10 hours later. On neurologic examination, optic fundi, visual acuity, visual fields, and pupil and eye movements were normal. Facial sensation and corneal reflex were symmetrical. A slight decrease in voluntary lower facial contraction was present on the right. There was no dysarthria or dysphagia, and the palate moved symmetrically. The gag reflex was normal. The tongue protruded in the midline. Tactile, pain, and temperature sensation were normal in the face and mouth. Tendon reflexes were symmetrical. The plantar reflexes were flexor. There was a slight distal paresis in the right hand, with decreased finger motility. There was a slight pronator drift with elevation of the right arm. There was a mild difficulty in reaching objects with the

right arm, without tremor or asterixis. There was no gait ataxia. Touch, pain, and temperature sensation were abolished in the right forearm. Position sensation of the fingers and wrist was abolished on the right. Stereognosis (recognition of small objects such as keys, coins, or a pin) and graphesthesia (recognition of figures drawn by the examiner on the palm of hand) were also abolished. Vibration (128 Hz) sensation was decreased (4/8) in the right wrist and hand. Neuropsychological examination with a standard battery of tests⁵ did not disclose any disturbance. General examination findings were normal. The patient's blood pressure was 150/90 mm Hg. The results of standard blood tests and urinalysis were normal except for an increased cholesterol level (7.3 mmol/L). The electrocardiogram was normal. Extracranial Doppler ultrasounds were normal. Computed tomographic scans of the brain on admission were normal. Magnetic resonance imaging performed after 6 days showed a hyperintense cortical and subcortical area in the left postcentral area at the level of the parietal operculum on T_2 -weighted images. The intensity was enhanced after gadolinium injection (Fig 1).

On discharge 11 days after admission, the findings of the neurologic examination had markedly improved. A slight decrease in pain and touch sensation still persisted in the fingers on the right, predominantly in the fourth and fifth fingers. Only a few disturbances in position sensation were still present, but severe stereognostic and graphesthesia disturbances persisted.

CASE 2.—On awakening one morning, a 71-year-old man experienced decrease of touch sensation in his left hand and numbness in the left side of his upper and lower lips.

He had untreated hypertension and insulin-independent diabetes. His right forearm had been amputated 22 years before as a result of a work injury. One year earlier, the patient had experienced transient weakness in the left upper limb. Computed tomography at that time showed a right frontal infarct.

On neurologic examination, the patient was alert and well orientated. Optic fundi,

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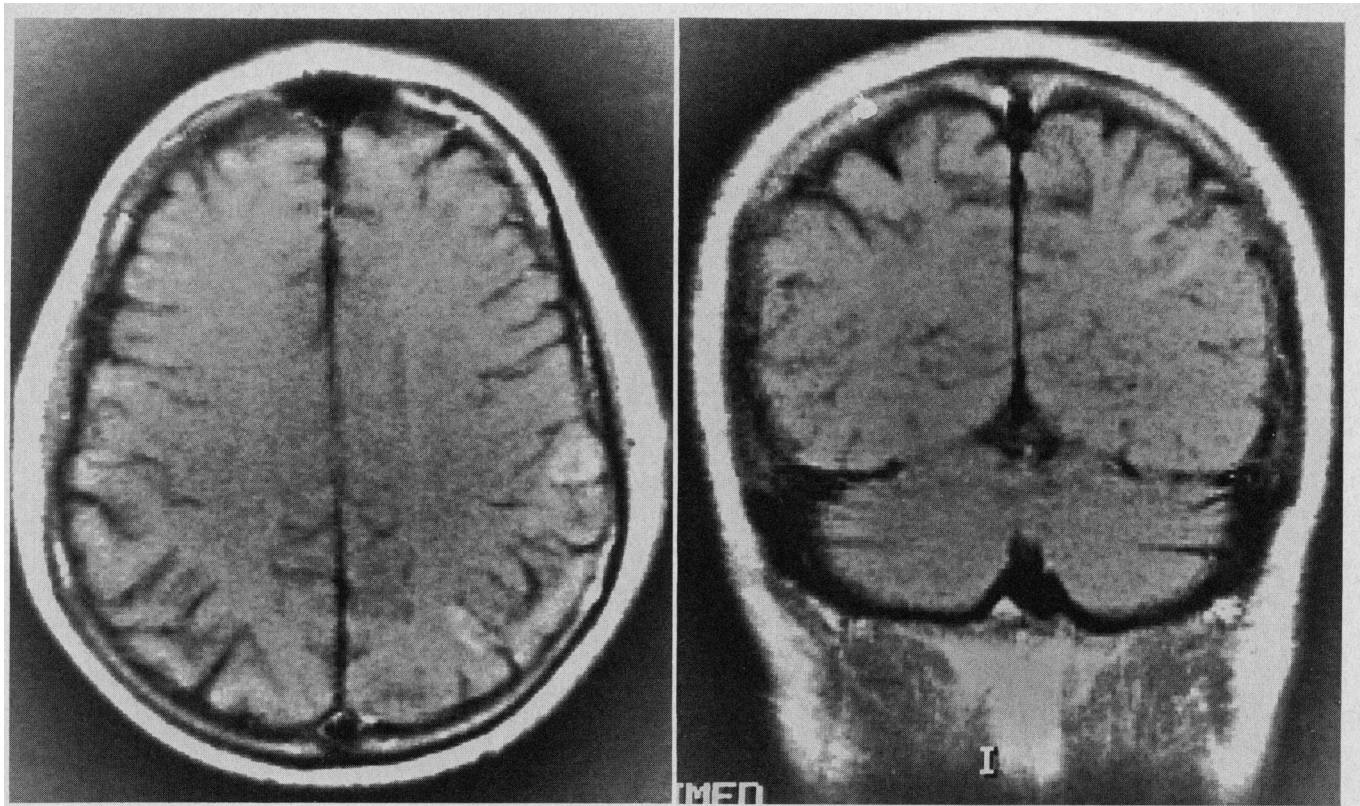


Fig 1.—Patient 1. Magnetic resonance T₁-weighted image after gadolinium injection. Transverse (left) and coronal (right) views show a postcentral infarct extending more caudally within the parietal lobe, involving mainly the cerebral cortex, with little white matter damage. The coronal section (right) shows involvement of the parietal operculum, with damage surrounding the sylvian fissure.

visual acuity, visual fields, and pupil and eye movements were normal. Facial sensation was preserved, with symmetrical corneal reflex. A slight lower facial paresis was present on the right. Tactile, temperature, and pain sensation were normal in the face and mouth. There was no dysphagia and the palate moved symmetrically. The gag reflex was normal. The tongue protruded in the midline. Tendon reflexes were normal and symmetrical. In the limbs, motor strength and tone were symmetrical and normal. There were elevation and abduction of the left arm, with flexion of the fingers. Fine movements of the fingers (piano playing) were decreased on the left. There was no dysdiadochokinesia or limb dysmetria.

Touch, pain, temperature, and vibration sensation were normal in the four limbs and trunk. Position sensation, stereognosis, and graphesthesia were nearly abolished in the left hand. Sensation was not decreased in the lower limbs, but the patient showed extinction of the left stimulus on simultaneous tactile stimulation of the lower limbs. The results of neuropsychological examination using a standard battery of tests⁸ were normal.

The findings of the general examination were normal except for a right carotid bruit. The patient's blood pressure was 219/115 mm Hg. The results of standard blood tests and urinalysis were normal except for a slight anemia (hemoglobin, 126 g/L), an increased cholesterol level (6.2 mmol/L), and slight hyperglycemia (glu-

cose, 6 mmol/L). The electrocardiogram was normal. Doppler ultrasounds and angiography showed a 90% stenosis of the right internal carotid artery. Computed tomography of the brain showed the old right frontal infarct and a recent right postcentral infarct at the level of the parietal operculum (Fig 2).

COMMENT

Both our patients developed cheiro-oral sensory dysfunction in relation to infarct involving the contralateral parietal opercular region. *Cheiro-oral* is a word with Greek-Latin origin, which describes subjective or objective sensory involvement in the peribuccal region and distal part of the upper extremities, mainly the hands and fingers.⁹ First described by Sittig⁷ in 1914, cheiro-oral sensory involvement, or cheiro-oral syndrome, was thought to be highly suggestive of a cortical lesion in the contralateral hemisphere and corresponded well to the sensory homunculus topography in the cerebral cortex.⁹ This assumption was mainly based on the development of paroxysmal cheiro-oral paresthesias, which were likely to be epileptic in patients with tumor or trauma. On the other hand, the role of thalamic involvement was subsequently empha-

sized in the development of cheiro-oral syndrome,⁶ and, in the literature, cheiro-oral syndrome resulting from supratentorial stroke has been reported only in thalamic infarction⁸⁻¹¹ or hemorrhage.^{11,12}

The first clinicopathologic study of cheiro-oral syndrome was carried out in 1917 by Schuster,¹³ who described a 50-year-old man who experienced paroxysmal tingling in his hand, fingers, and upper lip on the left side, with a slight decrease in position sensation on that side. At autopsy, an "endothelioma" (meningioma) was compressing the lower part of the postcentral gyrus and anterior part of the supramarginal gyrus on the right side; the brain parenchyma itself was not damaged. Seven clinicopathological studies were subsequently published, one involving another patient with a large parietal meningioma,¹⁴ two patients with multiple tuberculomas in the cerebral hemispheres,^{15,16} one patient with surgical lesion of the spinothalamic pathways in the midbrain,¹⁷ and three patients with lateral thalamic infarct limited to the ventral posterior nucleus.⁸⁻¹¹ In a review of the literature on cheiro-oral syndrome up to 1965, Hagenau⁶ could not find a

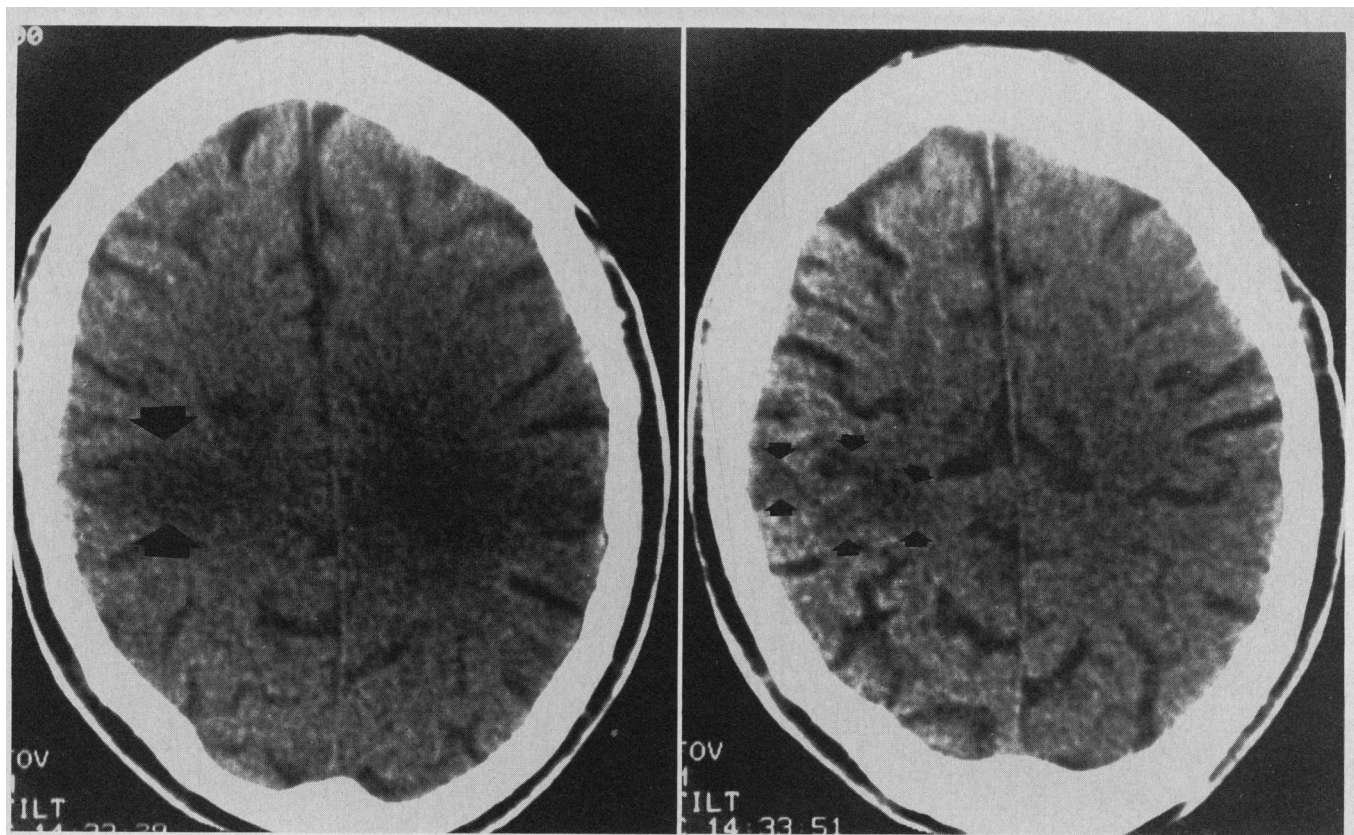


Fig 2.—Patient 2. Computed tomographic scans show a right postcentral infarct (arrows) extending into the underlying white matter.

single case of cortical stroke, and he emphasized thalamic involvement. To our knowledge, no patient has been described with cheiro-oral syndrome and stroke involving the cerebral cortex or underlying white matter on computed tomography or magnetic resonance imaging. Apart from thalamic infarction, cheiro-oral syndrome has been associated with pontine infarct,¹¹ hemorrhage,¹⁸⁻²¹ cavernous angioma,²² and midbrain hemorrhage²³ on computed tomographic and magnetic resonance imaging scans.

Sensory involvement in our patients was purely subjective in the face, but severe hypoesthesia was present in the hands, without any pseudoradicular distribution. This sensory deficit was "cortical" in type, as position sense, stereognosis, and graphesthesia were particularly severely involved (Vergier-Dejerine cortical hypesthesia^{24,25}). Pain and temperature sensation were involved in the first patient, but spared in the second patient. Vibration sensation was the least affected sensory modality in both patients. Involvement of the postcentral gyrus in the parietal operculum (somatosensory area I) may explain decreased position and discriminative touch sensation, while involvement of the more caudal portion of the parietal operculum and

insula (somatosensory area II) may explain decreased pain and temperature sensation (Dejerine-Mouzon hypoaesthesia²⁶) by damage to the thalamocortical pathways and to the projection area for nociception.²⁷⁻²⁹ Disturbed recognition (eg, stereognosis and pain perception) probably relates to more posterior involvement in the lower parietal region.²⁸ In fact, dissociated sensory disturbance involving proprioception, stereognosis, and graphesthesia may not be specific for cortical damage, as it has also been reported in pure sensory stroke secondary to thalamic infarction.²⁹

Another syndrome with hemisensory disturbance has been reported in patients with left anterior parietal artery territory infarction, associating conduction aphasia, mild motor impairment, and marked faciobrachiorural hypoaesthesia mainly involving the elementary modalities of sensation (ie, touch, position, pain, and temperature).^{30,35} In the single autopsied case, the infarct involved the postcentral gyrus P_1 and the anterior part of P_2 , but it also extended very deeply into the white matter down to the posterior limb of the internal capsule.³⁰ A pseudothalamic pain syndrome has also been reported in more posterior involvement of the parietal white

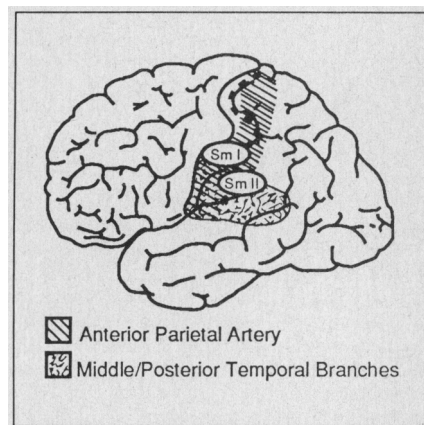


Fig 3.—Lateral view of cerebral hemisphere showing somatosensory areas I (Sm I) and II (Sm II) and arterial supply.

matter.³⁶ Involvement limited to the lower part of the postcentral gyrus, posterior parietal operculum, and underlying white matter in our patients may correspond to infarction in the territory of branches from both the anterior parietal artery and the middle and posterior temporal arteries, without more extensive damage to the anterior parietal artery territory (Fig 3). This type of partial superficial middle cerebral artery branch territory infarct seems rather uncommon, as

suggested by the absence of previous reports of cheiro-oral syndrome resulting from hemisphere stroke. This rarity is difficult to explain, but may be related to the vascular anatomy of the

posterior opercular region, which has a double vascular supply through branches from the temporal arteries and anterior parietal artery.^{37,41}

Our findings highlight the nonspec-

ificity of cheiro-oral sensory disturbance in acute stroke. They also provide evidence for partial middle cerebral artery territory infarction limited to the posterior opercular region.

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