

## Cheiro-Oral Syndrome

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### ABSTRACT

Cheiro-Oral Syndrome (COS) is a very rare neurological syndrome associated with varied etiology. We report a 53-year-old man presented with left sided perioral and ipsilateral hand/fingers burning sensation for a one-month duration. On examination, he had hypesthesia over left perioral and distal palmar aspect of all five fingers. MRI revealed subacute infarct in the posterior limb of right internal capsule adjacent to and minimally involving thalamus. He was diagnosed as Cheiro-Oral Syndrome as a result of ischemic stroke and managed.

### KEY WORDS

*Cheiro-oral syndrome, Stroke, Internal capsule, Perioral hypesthesia*

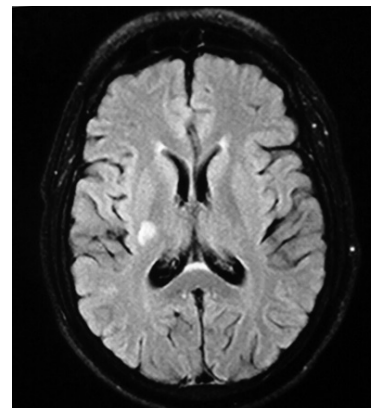
### INTRODUCTION

A sensory disturbance limited to the hand or fingers and the corner of the mouth both on the same side constitutes the cheiro-oral syndrome (COS).<sup>1</sup> It is a very rare neurological syndrome.<sup>2</sup> Although COS had received a great interest in Europe as a contralateral parietal lobe localizing value, with modern investigation technology the location of lesion producing COS ranges from the cortex to cervical spinal cord.<sup>3</sup> To our knowledge, this is the first case report of COS due to capsular infarct from Nepal.

### CASE REPORT

A 53 year old man had presented with burning sensation over left perioral and ipsilateral hand and fingers for a one-month duration. He had weakness of left side of the body and slurring of speech a week prior to the above symptoms. The weakness and slurring of speech gradually improved, but the abnormal perioral and hand sensations were persistent. He was a known hypertensive and diabetic on medication. He was a past smoker and consumed alcohol occasionally. Detail neurological and systemic examination

was normal except he had hypesthesia over left perioral and distal palmar aspect of all five fingers. MRI revealed subacute infarct in the posterior limb of the right internal capsule. (Figure 1)



**Figure 1.** MRI revealed sub-acute infarct in the posterior limb of right internal capsule adjacent to and minimally involving thalamus.

Laboratory investigations including CBC, LFT, RBS, RFT, HIV, HBSA/g, HCV, lipid profile and ECG and echocardiography were normal. We counseled him regarding the benign nature of his symptom, treated him with duloxetine for burning sensations, and initiated secondary prophylaxis for ischemic stroke.

## DISCUSSION

Although COS is a common neurological syndrome, it is mostly misdiagnosed. Since the first description by 1914 by Sir Sittig, a German doctor a number of studies on COS have been published.<sup>1,4-14</sup>

Although a simple definition had been put forward by Ten Holter and colleague,<sup>1</sup> WH Chen has tried to define it in detail so that the COS identification may be precise. As per this definition, COS is diagnosed if sensory disturbances are confined to the perioral area and finger(s)/hand without a detectable abnormality in mental, motor or cerebellar function.<sup>15</sup>

Interestingly our patient had all the features to be diagnosed as COS as per Chen's diagnostic criteria.

COS is divided into 4 types. Unilateral or Type I in which there is sensory impairment confined to the perioral area and ipsilateral finger(s)/hand. Bilateral or Type II in which there is sensory disturbance confined to the perioral area and finger(s)/hand bilaterally.<sup>16</sup> Atypically bilateral or Type III in which there is sensory disturbance confined to the perioral area and finger(s)/hand in that one is involved bilaterally whereas the other is unilateral.<sup>15</sup> Crossed or Type IV in which there is sensory disturbance confined to the perioral area and opposite finger(s)/hand in a crossed pattern.<sup>17</sup>

Our patient had Type I COS, which is a commonest variant (71.1%). Our patient's cause for COS was the ischemic stroke, which is known to be the leading cause for COS. In the study by Lin et al, the lesion for COS was the ischemic stroke (52.9%), followed by hemorrhagic stroke (21.8%).<sup>2</sup> Other causes included intracranial bypass complication (3.4%), cervical cord disorder (3.4%), neoplasm (2.9%), vascular malformation (1.1%), abscess, aneurysm, dermoid cyst, seizure, stereotactic surgery, middle cerebral artery stenosis, and drug (0.6%).<sup>15,18-26</sup> Almost 10% cases of COS were idiopathic.<sup>5</sup>

Our patient had a lesion in right internal capsule, which is known to be a rare cause for COS (approx. 4.0%). In the review by Lin et al, the most common location of lesions in COS was at thalamus (25.9%), followed by pons (24.7%), cortex (18.4%), internal capsule (4.0%), cervical cord (3.5%), corona radiata (2.9%), medulla oblongata (2.9%), midbrain (1.7%), and multiple sites (0.6%).<sup>2</sup> Although mostly in type I COS patients like ours, the lesion is found at thalamus to cortex, our case had capsular infarct.

The mechanism causing the peculiar distribution of sensory impairment in cheiro-oral syndrome resulting from a single lesion has been explained as a close somatotopic location in the postcentral gyrus of the parietal lobe.<sup>4,27-30</sup>

Clinicopathological correlation suggests that the sensory thalamocortical radiations lie farther posterior in the posterior limb of the internal capsule than the corticospinal motor fibers and that they probably lie adjacent to the thalamus.<sup>31</sup>

The explanation of the involvement of hand and mouth by the cortical lesion is difficult as upper part of the face is represented between these regions. These areas in the somatosensory cortex, like motor cortex, are thought to be so sensitive to stimuli that sensory impairment restricted to hand and corner of mouth is explained by a single lesion. Also, their fibers may be more vulnerable to injury.<sup>20</sup>

Although this can explain COS because of cortical lesion, it is postulated that thalamocortical fibers from hand are adjacent to those from the mouth in corona radiata.<sup>32</sup>

We feel that similar organization exists in the posterior limb of internal capsule, which explains our patient's manifestation.

The outcome in cases of COS in the literature is rarely mentioned. However, it is observed that as many as 16.5% of patient's neurological status deteriorate within 7 days after index COS.<sup>2</sup> In our case the patient already had symptoms for 1 month, the lesion was small and exclusively in the posterior limb of internal capsule, normal cardiac findings, and normal carotid doppler, we do not expect him to deteriorate in 6 months to come.

Although COS is a common neurological disorder, it is neglected and hence under-diagnosed or misdiagnosed. A capsular infarct is a rare cause for COS. The presence of this syndrome should alert a physician to localize a lesion from cortex to spinal cord so that the appropriate management can be offered.

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