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Cheiro-Oral Syndrome Secondary to Thalamic Infarction: A Case Report and Literature Review

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Abstract

Introduction—Small thalamic infarcts can present with a variety of sensory deficits that can be difficult to diagnose clinically because of their seemingly disconnected manifestations.

Case Report—Here we report a case of a 55 year old man presenting with the clinical findings of Cheiro-oral Syndrome, a pure sensory thalamic lacunar syndrome that clinically presents with contralateral sensory deficits of the fingers and hemi-mouth.

Conclusions—This report highlights both common and unusual clinical presentations of isolated thalamic infarctions, correlating clinical symptomatology with anatomic localization. In patients with such findings on exam, thalamic infarction should be considered, with a thorough workup initiated to both confirm the diagnosis and optimally reduce future stroke risk.

Keywords

Lacune; ischemic stroke; thalamus; Cheiro-Oral syndrome

Introduction

Cheiro-Oral Syndrome (COS) is a unique pure sensory thalamic lacunar syndrome that clinically presents with contralateral sensory deficits of the fingers and hemi-mouth. Cases of this syndrome have been reported in the literature secondary to both ischemic and hemorrhagic infarctions most commonly involving the thalamo-cortical projections, thalamus, or brainstem[1,2]. The syndrome can also be accompanied by deficits in other cranial nerves, the ipsilateral lower extremity, and/or including ipsilateral ataxia. Often the deficits include incomplete involvement of the face or extremities, making this diagnosis easily missed in the acute setting. Here we report a case of COS presenting acutely to our Emergency Department (ED) and discuss the implications of proper diagnosis and

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management. Other common and unusual thalamic infarction syndromes are also reviewed, relating the clinical presentation to the thalamic nuclei involved and the vascular supply to the thalamus.

Case Report

A 55 year old right-handed man with a previous medical history of hepatitis C, hypertension, 40 pack years of smoking, and asthma presented to our hospital ED with the complaint of mild numbness involving two digits of his left hand, the left corner of his mouth, and the left side of his tongue. The onset was sudden and without prodrome approximately 6 hours prior to ED arrival. Symptoms were persistent from onset through presentation. The patient denied recent illness, trauma, headache, confusion, mental impairment, or previous episodes of similar symptomatology. Blood pressure on admission was 104/69 and EKG demonstrated normal sinus rhythm. General physical and neurological examination were normal except subtle hypesthesia to temperature, light touch, and pinprick in the distal palmar aspect of the first 2 digits of the left hand, around the left corner of the mouth including the upper and lower lips, and on the left side of the tongue. No other cranial nerve deficits or sensory deficits were found. His strength, reflexes and gait were all normal. There was no ataxia, aphasia, apraxia, graphesthesia, or extinction. There were no laboratory abnormalities or acute changes seen on non-contrast head CT. Given the relatively minor clinical findings, coupled with the negative laboratory and head CT results, hospital admission versus discharge home from the ED with outpatient follow-up was debated. However, after a formal neurology consult the patient was admitted to the hospital for work up of a suspected lacunar infarct based on a clinical diagnosis of COS. An MRI performed shortly after admission showed an area of increased signal in the right posterior thalamus on FLAIR and diffusion weighted imaging (Figure 1). The corresponding region was dark on ADC sequences (not shown). The patient was diagnosed with a lacunar infarct in the posterior thalamus which was determined to be the cause of his sensory deficits. He was counseled about quitting smoking, and was started on both statin and anti-platelet therapies. Prior to discharge, two days after admission, the patient reported resolution of tongue numbness, however the sensory paresthesias in his fingers and ipsilateral peri-oral face persisted.

Discussion

Cheiro-Oral Syndrome is reported in the literature as having a number of variants, including lower extremity sensory loss, ataxic hemiparesis, and oculomotor deficits [3]. Reported causes for the syndrome include infarction of the cortex[4,5], operculum[6], corona radiata[7], posterior limb of the internal capsule[8], thalamus[9], or brainstem[10,11]. The presence of oculomotor (Cranial Nerve (CN) III) findings on exam serves to distinguish cases involving the brainstem[3]. Our case highlights the classic findings of isolated sensory deficits around the corner of the mouth as well as in the ipsilateral upper extremity; this is believed to occur secondary to the close mapping of these sensory homunculi regions in the thalamus[12,13]. The ventral posterior lateral (VPL) nucleus of the thalamus carries the sensory inputs from the upper extremity, while the ventral posteromedial (VPM) nucleus carries the trigeminal (CN V) sensory input. Tongue involvement occurs because general sensation of the anterior 2/3 of the tongue is supplied by the Lingual nerve which is a branch of CN V3. Because of the isolated and incomplete distribution of findings in this syndrome, it is tempting to place lacunar infarction lower on the differential diagnosis in the acute setting. Knowledge of the neuroanatomical basis for the seemingly disconnected mapping of symptoms is key to making the correct diagnosis. A history of sudden-onset symptoms in the setting of numerous vascular risk factors with the physical exam findings as described suggests the diagnosis and requires the initiation of a stroke work-up. In this particular case,

there was initial uncertainty in the ED about whether or not to admit the patient because the 'uncommon' clinical picture did not definitively suggest an ischemic infarction. Given the small lesion volume, and without the benefit of an MRI, this diagnosis may be suspected but cannot be confirmed.

COS is classified as a pure sensory lacunar syndrome with infarction in the VPM and VPL nuclei when thalamic involvement is implicated. There are many other common and rare unilateral thalamic lacunar syndromes, with the clinical symptoms depending on the size of the infarct, the specific thalamic nuclei involved, and whether adjacent structures are involved (Table 1). Simplistically, as seen in Figure 2 [adapted with permission from 13] and as listed in Table 2, the thalamus can be thought to contain 7 different nuclei including the ventral anterior (VA), ventral posterior (VP) - further divided into ventral posterior medial (VPM) and ventral posterior lateral (VPL), ventral lateral (VL), dorsomedial (DM), intralaminar (IL), pulvinar (P), and lateral geniculate body (LGB). Of note, inputs to the various nuclei of the thalamus are thought to be organized somatotopically[14]. Blood supply to the various thalamic nuclei occurs via small penetrating vessels that primarily originate from the posterior circulation vessels associated with the Circle of Willis. The larger parent vessels from which the small penetrating arteries typically originate include the basilar, posterior cerebral, and posterior communicating arteries. Figure 2 demonstrates this anatomically including some of the more common named penetrators including the tuberothalamic artery, the paramedian artery, the inferolateral artery, and the posterior choroidal artery. Table 2 further highlights the relationships between the various thalamic nuclei, the penetrating arteries, and the clinical presentation as related to the nuclei specific afferents and efferents. Here we also emphasize that there is considerable variability between the number of penetrators and their distribution to the thalamic territories supplied. Nonetheless, it is possible to localize thalamic infarctions using clinical presentation symptoms and a working knowledge of thalamic vascular anatomy.

Since the pathophysiology of lacunar infarction involves lipohyalinosis of the small penetrating arteries, management of risk factors is the most important step in preventing future infarctions. Complete work-up of this case allowed the diagnosis of an acute lacunar infarction to be made resulting in detailed patient counseling about his risk factors, especially smoking cessation, and the initiation of appropriate preventative statin and antiplatelet therapies. Diagnosing a stroke in this case may also make the patient more compliant with his long-term stroke preventative therapies, including lifestyle changes, and acts to further emphasize the importance of risk factor control to his medical caregivers. As such, close outpatient follow-up was initiated to optimally reduce his future stroke risk.

In conclusion, given the mild nature of the patient's disconnected hypesthesias it was tempting to write off the case as a mild peripheral or systemic process instead of a possible CNS infarction. The clinical suspicion of a lacunar syndrome based on history and physical exam resulted in the patient receiving a complete work-up and definitive diagnosis. Outpatient physicians, ED admitting officers, and neurologists should keep in mind the possibility that seemingly disconnected sensory deficits can indicate the presence of a small lacunar infarction.

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Figure 1.
FLAIR (A) and DWI (B) imaging demonstrating a right-sided thalamic lacunar infarct.

Figure 2.

Artist's rendition of major thalamic nuclei and arterial supply [adapted with permission from 13]. Schematic diagram of lateral (A) and dorsal (B) views of the 4 major thalamic arteries and the nuclei they supply. Numbers indicate the following: 1, carotid artery; 2, basilar artery; 3, P1 region of the posterior cerebral artery (mesencephalic artery); 4, posterior cerebral artery; 5, posterior communicating artery; 6, tuberothalamic artery; 7, paramedian artery; 8, inferolateral artery; and 9, posterior choroidal artery.

Table 1**Unilateral Thalamic Syndromes**

Thalamic Syndromes	Location of infarct	Presentation
Behavioral Syndromes [15] • Anterior Thalamus (dominant hemisphere) • Anterior Thalamus (non-dominant hemisphere)	AD, AM, AV, VA, rostral VL Left Anterior nuclei Right Anterior nuclei	Perseveration, confabulations, apathy and anterograde amnesia Verbal impairment Visuospatial impairment
Pure Sensory • Cherie Oral Syndrome • Dejerine-Roussy Syndrome [16]	VPM (head), VPL (trunk& extremities) VPM, VPL VPM, VPL, VL	Broad: Contralateral paresthesia or dysethesia or other abnormal sensation Contralateral hemioral and hand Contralateral hemibody pain
Mixed Sensorimotor	Thalamus & adjacent posterior internal capsule	Broad: Contralateral hemiparesis and sensory impairment
Ataxic Hemiparesis • Hemiataxia-hypaesthesia [17] • Hypaesthetic ataxic hemiparesis [17]	Inferolateral Thalamus VL, VP VL, VP, PLIC	Broad: Contralateral ataxia and hemiparesis Contralateral ataxia & sensory impairment Contralateral ataxia, sensory impairment, & motor weakness

AD - Anterior Dorsal nucleus; AM - Anteromedial nucleus; AV - anteroventral nucleus; VA - Ventral Anterior nucleus; VL - Ventral Lateral nucleus; VPM - Ventral Posteromedial nucleus; VPL - Ventral Posterolateral nucleus; VP - Ventral Posterior nucleus; PLIC - Posterior Limb of Internal Capsule.

Table 2**Major Thalamic Nuclei**

Nuclei	Afferent Projections	Efferent Projections	Proposed Function	Arterial Supply
VA	Substantia nigra, internal globus pallidus, deep cerebellar nuclei	Prefrontal, premotor, motor & supplementary motor cortex	Relays basal ganglia to cerebellar inputs to cortex	Tuberothalamic (arises from Posterior Communicating Artery)
VL	Internal globus pallidus, deep cerebellar nuclei, substantia nigra	Motor, premotor, supplementary motor cortex	Relays basal ganglia and cerebellar inputs to cortex	Paramedian (arises from P1), Tuberothalamic, Inferolateral (arises from P2)
VP - VPL	Medial Lemniscus, spinothalamic tract	Somatosensory cortex	Relays somatosensory spinal inputs to cortex	Inferolateral
VP - VPM	Trigeminothalamic tract, taste inputs	Somatosensory and taste cortex	Relays somatosensory cranial nerve inputs and taste to cortex	Inferolateral
DM	Amygdala, olfactory cortex, limbic basal ganglia	Frontal Cortex	Limbic pathways, major relay to frontal cortex	Paramedian, Tuberothalamic
IL	Deep cerebellar nuclei, globus pallidus, brainstem, ARAS, sensory pathways	Cerebral cortex, striatum	Maintain alert consciousness, motor relay for basal ganglia and cerebellum	Paramedian
P	Tectum, other sensory inputs	Parietotemporo-occipital association	Behavioral orientation towards relevant visual & other stimuli	Posterior Choroidal (arises from P2)
LGN	Retina	Primary visual cortex	Relays visual inputs to cortex	Posterior Choroidal

VA - Ventral Anterior nucleus; VL - Ventrolateral nucleus; VP - Ventral Posterior nucleus; VPL - Ventral Posterolateral nucleus; VPM - Ventral Posteromedial nucleus; DM - Dorsal Medial nucleus; IL - Intralaminar nuclei; P - Pulvinar nucleus; LGN - Lateral Geniculate Nucleus; P1 & P2 - segments of Posterior Cerebral Artery; ARAS - Ascending Reticular Activating Systems.