Restricted Acral Sensory Syndrome
Following Minor Stroke
Further Observation With Special Reference to Differential Severity of Symptoms Among Individual Digits

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Background Restricted acral sensory syndrome (RASS) after minor stroke most often manifests as a cheiro-oral syndrome. However, recent studies have described more varied patterns of RASS and also have reported that the degree of sensory symptoms may vary among individual digits. Until recently, however, there have been no reports in which sufficient numbers of patients were studied with detailed information on the symptomatic severity among individual digits.

Summary of Review In this report, I describe 30 patients presenting with RASS secondary to minor stroke. Computed tomographic scan and/or magnetic resonance imaging identified lesions in the lateral thalamus in 11, midbrain in 2, pontine tegmentum in 8, capsulo-corona radiata in 5, and frontoparietal subcortical-cortical areas in 4 patients. The patterns of RASS were cheiro-oral in 10, cheiro-oral-pedal in 8, cheiro-pedal in 4, restricted to palm and/or fingers in 7, and periotal-pedal in 1. Dominant involvement of upper lip, thumb, and index finger was frequent, especially in patients with thalamic and thalamocortical lesions. In patients with cortical-subcortical lesions, cheiro-oral or restricted finger involvements were observed, while the foot was spared. In patients with pontine lesions, bilateral RASS was occasionally observed, and the pattern of preponderant involvement of the first two digits was not apparent.

Conclusions These patterns of RASS generally agree with the previously observed sensory topography of monkeys, and they support anatomic proximity of sensory fibers from acral parts of the body. However, other mechanisms such as differential vulnerability of generation of paresthesia in different body parts or a low-threshold concept based on disproportionately large representing areas for the acral parts of the body in the human sensory system may also be required to explain some of the clinical observations. (Stroke. 1994;25:2497-2502.)

Key Words • cerebrovascular disorders • motor activity • neurons, sensory

Sensory abnormalities after minor stroke are occasionally restricted to the acral part of the body.1 This restricted acral sensory syndrome (RASS) most often presents as a pattern of cheiro-oral syndrome (COS), a combined sensory abnormality over the perioral and ipsilateral palm area.2-23 Although COS was first reported in patients with parietal lesions,2 minor strokes involving the thalami,3,4,6-9,12,14,16-18,21 brain stem,5,10,11,13,15,16,18-20 and corona radiata23-25 have subsequently been reported to produce this syndrome. More recently, patients with restricted sensory changes in the palm and foot (cheiro-pedal syndrome [CPS]),26,27 or in the palm, foot, and perioral area (cheiro-oral-pedal syndrome [COPS])27,30 were reported, although a similar pattern of sensory abnormality was already described in earlier studies of Fisher.31-32 Furthermore, in some of the patients with RASS, differential severity of sensory symptoms among digits was often identified.1 For instance, some patients were reported to have limited sensory changes in the first two or three fingers.4-5,15,21

These observations have evoked speculations and controversies regarding the topography of the sensory pathway in the human nervous system. Despite numerous case reports, however, there have been no studies in which sufficient numbers of patients were described with detailed information on differential symptomatic severity among individual digits. In this report, I review my personal experience of 30 patients who developed various patterns of RASS secondary to minor stroke with special reference to differential severity of paresthesia among digits.

Subjects and Methods
At Asan Medical Center, I examined 36 patients who developed RASS after minor stroke from January 1989 to August 1994. Thirty patients showed lesions in the computed tomographic (CT) scans and/or magnetic resonance imaging (MRI) that were considered appropriate to the clinical findings, whereas 6 with negative or equivocal radiological findings were excluded. Twenty-one of 30 patients showed sensory disturbances restricted to the acral parts when they were first examined; in the 9 remaining patients, the sensory abnormality was initially more generalized (such as diffuse sensory change in the forearm or lower leg), but it had become restricted to the acral parts by follow-up examination, usually within several days. Separation of these two groups was not attempted because patients showing RASS on initial examination might also have had more diffuse sensory deficit had they been examined earlier. All patients were examined by the author, with detailed sensory testings at each examination.
Results

Thirty patients (15 men and 15 women, aged 37 to 88 years [mean, 58 years]) were studied. Twenty-five had infarcts and 5 had small hemorrhages. Risk factors for vascular disease included hypertension in 25, diabetes mellitus in 6, cigarette smoking in 2, and cardiac disease in 2. Among 3 patients without conventional vascular risk factors, 2 (patients 13 and 24) showed positive lupus anticoagulant tests, and 1 (patient 28) with pontine hemorrhage was found to have vascular malformation at surgery. In 14 patients, sensory disturbances were the only clinical manifestations (pure sensory stroke), whereas in others minor nonsensory neurological disturbances were noted.

All patients complained of acral paresthesia. In 14 patients, there were no objective sensory disturbances. In others, sensory deficit of pinprick, vibration, touch, and position was noted, which was usually mild, tended to fluctuate or change on each examination, and improved rapidly. The area of objective sensory deficit was usually identical to that of paresthesia, although some patients initially presented with decreased pinprick and/or vibration sense in a wider area. The paresthesia over the acral parts of the patients’ bodies was distinct and relatively persistent and thus used in this clinical-topographical correlation study. The sensory topography was drawn in Figs 1 through 4 according to the symptomatic severity (severe, black; moderate, dense dots; mild, sparse dots). The patterns of RASS included COS in 10, COPS in 8, CPS in 4, restricted to palm and/or fingers in 7, and periotal-pedal in one. In all patients, the acral paresthesia persisted in the same location at least for 1 week, usually for several months. In most cases, however, the severity of paresthesia gradually diminished, and the differential degree of severity among digits became blurred. However, in a few patients (Nos. 1, 2, and 23), uncomfortable and often burning acral paresthesia persisted for more than 6 months. To rule out the presence of peripheral
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Restricted Sensory Syndrome

Pt / Sex / Age  Sensory topography  Other neurologic signs  Lesion location

21 / M / 63  3rd nerve palsy
22 / F / 44  Ataxia
23 / M / 56  Hemiparesis  One - and - a - half syndrome
24 / F / 39  Quadriparesis
25 / M / 67
26 / M / 56
27 / M / 67  6th nerve palsy
28 / F / 37  Ataxia  Gaze paresis
29 / M / 88
30 / M / 66  INO  Quadriparesis

Fig 1. Sensory topography of patients with thalamic stroke. Ten patients had infarcts, and 1 (patient 11) had a hemorrhage in the lateral thalamus, probably involving the nucleus ventralis posterior (VP); they showed various patterns of RASS: COS in 5, COPS in 5, and symptoms restricted to fingers in 1. Among 10 patients with perioral involvement, 8 had symptoms restricted to the area around the upper lip, and 2 had symptoms in upper and lower lips as well as intraoral areas. All patients had paresthesia in the fingers; 4 had numbness in the palm as well. However, dorsum of the hand was spared in all. The severity of paresthesia was equal among fingers in 3 patients, while 8 had symptoms more marked in the fingers on the radial side; in 2 patients (Nos. 4 and 7), paresthesia was restricted to the tips of the thumb and the index finger. Paresthesia of the foot and toes, noted in 5 patients, was generally mild compared with the symptoms of lips or fingers; 2 patients (Nos. 2 and 10) showed preponderant symptoms in the toes on the medial side.

Fig 5. Computed tomographic scan shows a small hemorrhage (arrowhead) in the left lateral thalamus that produced chiro-oral syndrome (patient 11).

Corona Radiata or Internal Capsular Strokes
Sensory topography of these patients is shown in Fig 2. Three patients (Nos. 12, 15, and 16) with corona radiata infarct presented with COPS, CPS, and COS, respectively, and 2 with lenticulocapsular infarct presented with CPS. Patient 12 had paresthesia limited to the first three fingers and toes, patients 13 and 15 had diffuse symptoms in the palm and foot, and patient 14 had paresthesia limited to the tips of the volar side of the first and second fingers and the big toe.

Frontoparietal Subcortical or Superficial Cortical Strokes
One patient with frontoparietal subcortical infarct, 1 with frontal subcortical infarct, and 2 with frontoparietal superficial infarcts were observed (Fig 3). Patient 17 had COS with equal symptom severity among fingers, whereas patients 20 and 18 showed paresthesia limited to the hand or the tips of the first four fingers, respectively. Patient 19 with discrete cortical infarct presented with persistent paresthesia limited to the tips of the third and fourth fingers.

Brain Stem Strokes
Ten patients were identified (Fig 4). Of the 2 with midbrain infarct, patient 21 presented with COS with equal severity among the fingers, while patient 22 had...
symptoms limited to the palm and fingers, which were more marked in the first two fingers. Eight patients with pontine stroke showed diverse patterns of RASS. Patients 23 and 28 showed bilateral COPS, and patient 26 had COS of bilateral perioral and unilateral palmar involvement. Patient 29 had symptoms limited to the contralateral upper lip and ipsilateral fifth finger. Patients 25 and 30 had symptoms restricted to fingers, with the preponderant severity in the middle three fingers. Patient 24 had two consecutive infarcts in the pons and presented with bilateral CPS. Patient 27, with isolated sixth nerve palsy, had paresthesia in the lateral face (perioral area) and the sole of the foot. The severity of paresthesia was more marked in ulnar-side fingers of both hands of patient 24 and in one hand of patient 28. In the 4 patients with foot involvement, the severity of paresthesia was generally equal among toes.

**Discussion**

Fisher,1 after analyzing 135 patients with pure sensory stroke, stated that isolated paresthesia of the face, arm, and leg suggests thalamic involvement, whereas selective involvement of some, rather than all, fingers may indicate a cortical localization. Brain stem lesions were not considered to follow any of these patterns. This assumption, however, was not based on pathological or radiological data. Our 30 patients with RASS demonstrated that various patterns of RASS can be caused by strokes involving any portion of the human sensory pathway.

Of the 11 patients with thalamic stroke, 5 had COS, 5 had COPS, and 1 had limited sensory dysfunction in the fingers. Eight of 11 patients showed dominant paresthesia in the radial-side fingers as observed in previous reports,1,4,6,21 and 2 (patients 2 and 10) of 5 with foot involvement showed symptoms predominantly in the medial toes. Restricted paresthesia of the medial two toes was previously described in a patient with pathologically proven thalamic lacune.1 Among 10 patients with perioral involvement, 8 had symptoms limited to the upper part of the lip suggesting that, like digits, the perioral area may not be evenly affected in patients with RASS.

According to previous studies using macaque monkeys,33-35 sensory projection fibers from the face, hand, and foot are arranged from the medial to lateral side in the ventral portion of the VP, where the tips of individual digits occupy large representation areas. The projection areas from proximal limbs and the trunk are relatively small and located in the dorsal area. In the hand representation area, the area for the thumb is located the most medially, and that for the little finger is located most laterally. Similar topography was also suggested in the study of humans.36 In the topographic scheme of monkeys, the areas for the thumb and the index finger lie in the same rostrocaudal plane and are adjacent to the upper lip area, which is disproportionately large in the face representation area (Fig 6).35 This topography, if applied to humans, may explain the preponderant involvement of the first two fingers and the upper part of the lip. Furthermore, the toe representation area is adjacent to the finger areas, with the area for the big toe situated most medially and that for the little toe most laterally (Fig 6). Therefore, a sensory pattern of COPS or CPS is possible without involvement of the trunk or proximal limbs. The preponderant involvement of the volar rather than dorsal surface of the hand or fingers in our patients may also be consistent with their topographic scheme, in which projection areas from the dorsum of the hand are located dorsally, separated from those of digits.

The sensory topography is less well characterized in the thalamocortical tract in the internal capsule or corona radiata.25,37 Corona radiata strokes causing COS have been reported by previous authors23-25 who believed that sensory fibers from perioral and hand areas may also run in close proximity in this area. Our 2 patients with striatocapsular infarcts and 1 with corona radiata lesion presented with CPS, and 1 patient with corona radiata lesion showed COPS, illustrating that COS is not the only pattern of RASS caused by lesions involving this area. The patients described by Derosnesé et al26 and Yasuda et al30 with corona radiata strokes also presented with CPS and COPS, respectively. In our study, patients 12 and 14 showed more severe symptoms in the radial fingers and medial toes. Two of the 7 patients with COS in recent reports34,35 also had selective involvement of the first two fingers. Therefore, the sensory topography in the thalamocortical pathway may be similar to that of the VP of the thalamus, as suggested by previous studies using macaque monkeys.38

I observed 4 patients with frontoparietal subcortical or cortical infarcts. The absence of foot involvement in this group suggests that the sensory fibers from the foot are separated below this level and agrees with previous demonstrations that the area representing the foot is located apart from finger areas in the parietal cortex in contrast to the thalamus.33,38,39 The findings from patient 17 of this study and previously reported patients with COS secondary to cortical lesions30 also support the anatomic proximity of sensory fibers from the perioral and finger areas in the cortex39-43; these findings are most consistent with the scheme of primates drawn by Woolsey in which the representation areas for lip and fingers are adjacent to the central sulcus, and the upper face is represented more posteriorly. However, patients 18 through 20 presented with sensory changes restricted to the palm or some of the fingers, suggesting that the lesions involved a portion of widely dispersed but somatotopically arranged sensory fibers in the superficial cortical area. Of interest was patient 19, who presented with paresthesia limited to the tips of the third and fourth fingers. This case is in line with a recent report of
small cortical infarct producing selective impairment of the motor power of the thumb, and it matches perfectly with previous topography illustrating that representation areas for the third and fourth fingers are adjacent in the parietal cortex. Isolated sensory changes in the hand, first four fingers, or second and third fingers have been previously reported in patients with parietal cortical lesions.

In the pons, the sensory fibers from the mouth, arm, and leg are located from the medial to lateral side in the small area of the medial lemniscus. Therefore, bilateral sensory deficit, especially paresthesia of the body, can be caused by mid-pontine lesions, as in our patients 23, 26, and 28 and in previously reported cases. In our patients, the severity of symptoms among individual digits was diverse, and the pattern of preponderant involvement of the first two digits was not apparent. However, a few patients with pontine stroke were described as having dominant symptoms in radial fingers. The precise sensory topography of digits remains unclear for the brain stem. According to previous studies using monkeys, the thumb is projected most laterally and the little finger most medially in the parietal cortex. Isolated sensory changes from acral parts of the body. However, this alone may not be sufficient to explain the sparing of the trunk, upper face, and proximal limbs, especially in patients with relatively large pontine lesions (patients 23 and 28) that probably involved sensory fibers from wide areas of the body. Furthermore, in several topographical studies, areas representing the forelimb and hindlimb were suggested to be located between areas for fingers and toes in the thalamus, and the area representing the upper face between areas for lip and fingers in the parietal cortex, although it is not clear whether these are located in the same plane as the acral parts of the body. The anatomic-proximity concept also cannot explain why big toes were more severely involved than ulnar-side fingers in patients 2, 10, 12, and 14. Therefore, the low-threshold (and consequent high-vulnerability) concept based on disproportionately large representation areas for acral parts in the human sensory pathway seems to be required to explain these observations. A lower sensory threshold of fingers and toes was previously documented in an experiment using baboons, and a recent study reported that representation areas for individual digits are different in size in the human parietal cortex.

In addition, the acral areas where the mechanoreceptors have small receptive fields with strong surround inhibition at multiple levels of sensory processing may be more vulnerable for perturbation of inhibitory-facilitatory balance and thus be more apt to produce paresthesia. Finally, the presence of an uncrossed, polysynaptic ascending spinal cord sensory pathway from the trunk and proximal limbs may, at least in part, contribute to the preferential sparing of these areas in patients with unilateral lesions. These mechanisms in combination with anatomic-proximity and low-threshold concepts provide the basis for the understanding of the patterns of RASS reported here.

Acknowledgments

I wish to thank Jun Kim, MD, PhD, and S. Baik, MS, for valuable discussion and S. Yoon, RN, for the help in preparing the manuscript. Some of the patients reported here were included in my previous articles.

References


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Stroke. 1994;25:2497-2502
doi: 10.1161/01.STR.25.12.2497

Stroke is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1994 American Heart Association, Inc. All rights reserved.
Print ISSN: 0039-2499. Online ISSN: 1524-4628

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://stroke.ahajournals.org/content/25/12/2497

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