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CASE REPORT

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Short title

Superficial Siderosis with CBS

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Abstract

A 70-year-old man presented with slow gait and a clumsy hand on the right side. Neurological examination revealed dystonia and ideomotor apraxia in the right hand. A mask-like face, rigidity in the right arm, and shuffling gait were noticed. Treatment with L-dopa had no effect. He was clinically diagnosed with corticobasal syndrome (CBS). Susceptibility-weighted images of brain magnetic resonance imaging showed low-intensity areas on the surface of the cerebral cortex, especially on the left parietal lobe, brain stem, and cerebellum, indicating superficial siderosis (SS). Neuropathological examination revealed diffuse hemosiderin deposition over the surface of the brain, including the left parietal lobe. Cerebellar hemorrhage with rupture to the subarachnoid space was noticed. Cortical SS should therefore be included in the differential diagnosis of CBS.

KEYWORDS

dystonia; hemosiderin deposition; ideomotor apraxia; susceptibility-weighted image

1 INTRODUCTION

In addition to corticobasal degeneration (CBD), the background pathology of corticobasal syndrome (CBS) includes a wide variety of diseases, including Alzheimer's disease, progressive supranuclear palsy, frontotemporal degeneration, Lewy body disease, and even Creutzfeldt–Jakob disease[1]. Here, we report a case of probable CBS caused by cortical superficial siderosis (SS).

2 CASE REPORT

A 70-year-old man first noticed fatigue with less daily activity. Mild cognitive decline with mini-mental state examination (MMSE) 27/30 points was noted at a nearby clinic. One year later, he gradually developed clumsy hand on the right and was referred to our hospital. Neurological examination revealed further cognitive decline with MMSE 23. Ideomotor apraxia with uncontrollable dystonic groping hand movement (alien limb phenomenon) was noticed dominantly on the right. He had a mask-like face, rigidity in the right arm, and shuffling gait without muscle weakness, ataxia nor abnormality in tendon reflexes. Treatment with L-dopa 300 mg for 11 months had no effect. He was clinically diagnosed with probable CBS fulfilling the most recent criteria[2].

T2-weighted brain magnetic resonance imaging (MRI) images showed no abnormalities including lateralized cortical atrophy (Figure 1A). Susceptibility-weighted imaging (SWI) showed a low-intensity area diffusely on the surface of the cerebral cortex, especially around the left Sylvian fissure as well as the brain stem and cerebellum, indicating SS (Figure 1B, C). Subcortical low-intensity spotty lesions, compatible with cerebral microbleeds, and cerebellar hemorrhages were also noticed in the SWI. Cerebral blood flow (CBF), evaluated with single photon computed tomography and rated with z-score, was projected on the cerebral cortex using 3D mapping. A marked decrease in CBF was noticed in the lateral parietal lobe and upper temporal lobe on the left (Figure 1D) as well as in the left

basal ganglia.

He developed hearing disturbance at the age of 73. His cognitive function and gait deteriorated gradually, and he died at 78.

Gross neuropathological examination revealed brown pigmentation on the cerebellum, orbital surface of frontal lobe, temporal lobe, and olfactory bulb (Figure 1E). A large number of hemosiderin-laden macrophages with gliosis were seen in the left orbital surface of the frontal lobe and from the temporal lobe to the angular gyrus (Figure 1F). The affected cortices were accompanied by leptomeningeal hemosiderin deposition. Chronic cerebellar hemorrhage extended from the dentate nucleus to the white matter with rupture to the subarachnoid space (Figure 1G). No vascular abnormality was found around the hemorrhage. Lipohyalinosis in the leptomeningeal vessels with hemosiderin deposition was occasionally observed (Figure 1H). Frame-shaped neurofibrillary tangles were restricted to the entorhinal cortex, with classic plaques rarely observed in the neocortex. Amyloid β immunopositive leptomeningeal vessels were occasionally observed. Ballooned neurons, astrocytic plaques, tuft-shaped astrocytes and globose-type neurofibrillary tangles were not observed.

3 DISCUSSION

The present case developed right-dominant motor and cognitive symptoms. The asymmetric presentation with limb rigidity plus akinesia and dystonia (2 of 3 motor features) as well as limb apraxia and alien limb phenomena (2 of 3 cortical dysfunctions) fulfills the criteria of probable CBS[2].

In the present case, autopsy identified SS as the cause of CBS. In addition to CBD, a wide variety of diseases have been reported as causes of CBS[1]. Vascular diseases, mostly

ischemic, may even mimic CBS[3]. The distribution of the lesions, including cerebral cortex and basal ganglia, commonly affected in these cases and the present case may explain the apparently similar clinical features. Neuropathological examination excluded the possibility of CBD, Alzheimer's disease, Parkinson's disease, and progressive supranuclear palsy, which may present with CBS. The present report is the first to show that asymmetric SS can cause CBS with contralateral dominance, aside from a report of SS with CBS that showed no asymmetry in SS[4]. Hemosiderin, released from the cerebellar hemorrhage with subarachnoid rupture, conveyed and deposited on the cerebral cortex around Sylvian fissure is regarded as the cause of SS in the present case.

In conclusion, we have presented the first case of SS presenting with probable CBS with asymmetric features. Cortical SS should be included in the differential diagnosis of CBS.

ACKNOWLEDGEMENTS

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CONFLICT OF INTEREST

The authors declare no conflicts of interest for this article.

CONSENT STATEMENT

Informed consent was obtained from the family for publication of this case.

Figure legends

Figure 1. Brain magnetic resonance imaging (MRI) (A-C), single photon emission computed tomography (D) and pathological images (E-H). A; T2-weighted image showed no apparent lesion. B and C; Susceptibility-weighted images showed diffuse cortical superficial siderosis, especially around the left Sylvian fissure (arrowhead). D; A marked decrease in cerebral blood flow was noticed in the left lateral parietal lobe and upper temporal lobe. E; Brown pigmentation was dominantly found around the left Sylvian fissure (arrowheads). Bar 1 cm. F; A large number of hemosiderin-laden macrophages with gliosis were seen in the superficial layer of the left angular gyrus. Berlin blue stain, bar 100 μm. G; Chronic cerebellar hemorrhage extended from the dentate nucleus to the white matter (arrow), with subarachnoid rupture (arrowheads). Bar 1 cm. H; Lipohyalinosis of the leptomeningeal artery was occasionally observed with hemosiderin deposition. Hematoxylin eosin stain, bar 50 μm.

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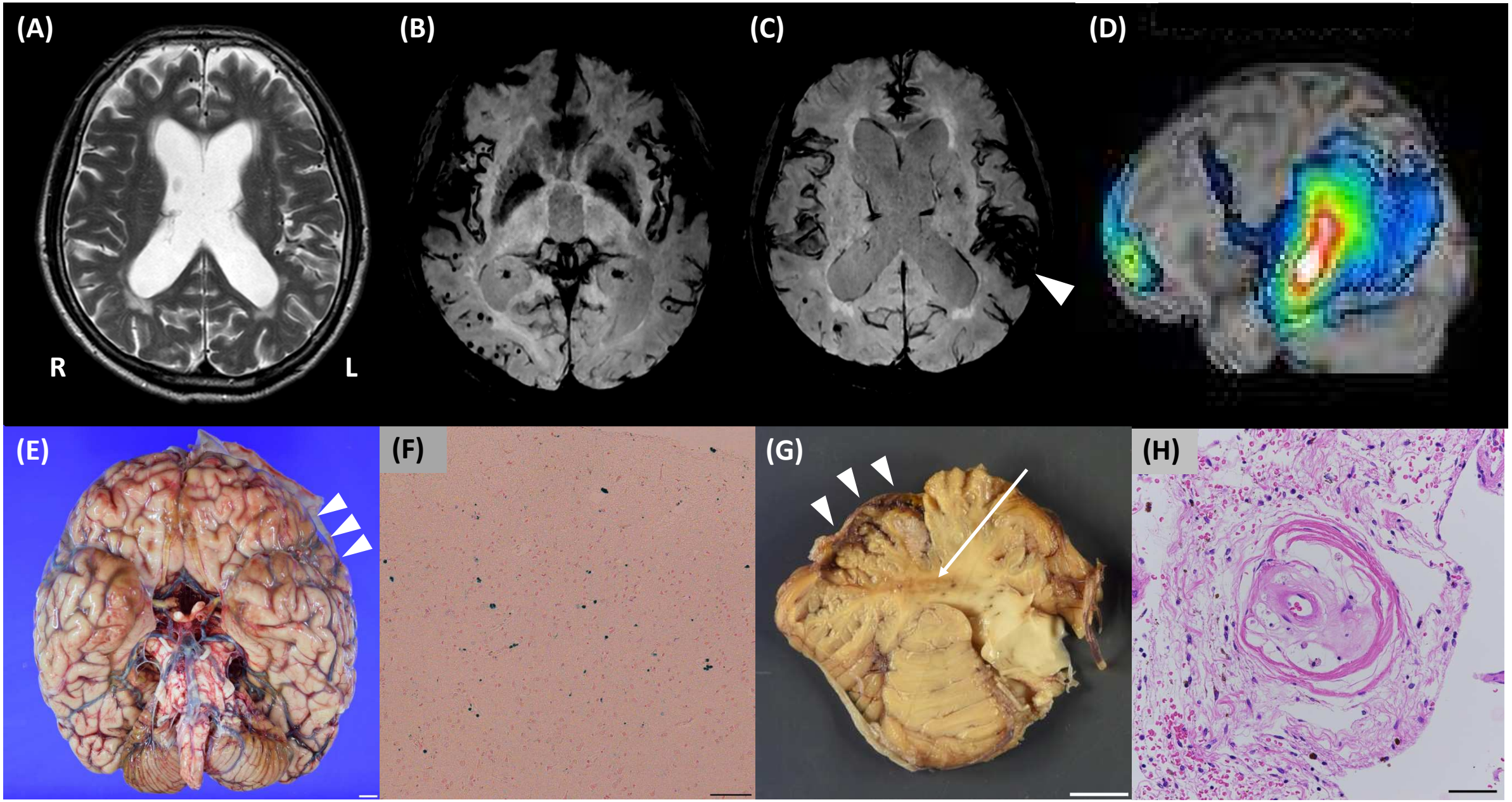
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