## Case Report

# Malignant Middle Cerebral Artery Infarction Resulting from Dissection of Middle Cerebral Artery

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A 30-year-old man presented with headache, left hemiparesis, and uncal herniation. Magnetic resonance imaging showed acute infarction in the right middle cerebral artery (MCA). Lifesaving craniotomy proceeded because of the uncal herniation. Assessment of brain tissue led to a diagnosis of MCA dissection. His skin biopsy showed ultrastructural abnormalities of dermal connective tissue. This is the unique case that dissection was diagnosed from both a biopsy specimen of the MCA and the dermal connective tissue. **Key Words:** Middle cerebral artery—dissection—malignant middle cerebral artery infarction—skin biopsy—acute ischemic stroke. © 2015 by National Stroke Association

Arterial dissection is relatively rare in the anterior compared with the posterior circulation, and middle cerebral artery dissection (MCAD) is exceptional, with only a few cases having been reported,<sup>1-4</sup> particularly in Japanese.<sup>5</sup> Herein, we describe a Japanese patient with rapidly progressive ischemic stroke caused by right MCAD who underwent craniotomy for decompression. Neuropathologic findings revealed right MCAD.

### **Case Report**

A 30-year-old man with moderate dyslipidemia was admitted to our hospital with left-hand clumsiness. Two days before admission, he developed a headache with

There are no disclosures.

drug resistance. He did not have history of smoking, problem alcohol drinking, or drug abuse. He had no familial history especially in collagen diseases. Physical examination did not reveal hyperextensible skin or moles. Neurologic examination revealed drowsiness, right conjugate deviation of the eyes, left hemispatial neglect, and left hemiparesis including the left side of the face. The score on the National Institutes of Health Stroke Scale was 11. Antinuclear antibody and autoantibodies were negative, and alpha 1-antitrypsin and total homocysteine levels were normal. He did not have mutation in *COL3A1*.

Brain magnetic resonance (MR) with diffusionweighted imaging immediately after admission showed focal acute infarction in the right middle cerebral artery (MCA) area (Fig 1, A,E), and MR angiography (MRA) showed stenosis in the top of right internal carotid artery and in the right MCA M1 and M2 segments. No embolic sources were detected by transthoracic echocardiography or conventional carotid ultrasonography.

His clumsiness gradually worsened after admission. On hospital day 4, his left extremities became flaccid, and the headache in his right temporal region continued. Brain MR imaging and MRA showed enlargement of the infarction in diffusion-weighted imaging, and the right internal carotid artery was abnormal compared with the left (Fig 1, B,F). We initially considered a differential diagnosis of reversible cerebral vasoconstriction syndrome, primary

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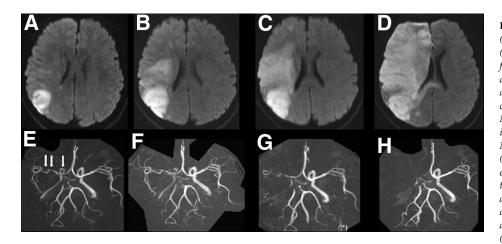


Figure **1.** Magnetic resonance (MR) with diffusion-weighted imaging (DWI) and MR angiography (MRA) findings from admission until hospital day 9. On admission day, DWI shows acute focal infarction in right middle cerebral artery (MCA) area (A), and MRA shows stenosis at top of right internal carotid artery (ICA) and right MCA M1 and M2 segments (arrow) (E). Image on hospital day 4 shows enlarged infarction in DWI (B), and that of right compared with left ICA is abnormal (F). DWI images on hospital day 6 show enlarged infarction (C), and right ICA image has worsened (G). Images on hospital day 9 show totally occluded right ICA-MCA (D) and herniation (H).

angiitis of the central nervous system, and arterial dissection. However, intravenous methylprednisolone therapy did not improve the symptoms, and the infarct region expanded (Fig 1, C,G). He fell into a coma with anisocoria and total occlusion of the right ICA–MCA (Fig 1, D,H) on hospital day 9. We performed a lifesaving craniotomy for decompression. Biopsy specimens of the right frontotemporal lobe showed dissection at the M2 and M3 segments of the right MCA (Fig 2, A,B). Neuropathologic findings revealed accumulated red blood cells above the internal elastic lamina without collapse of the lamina, and increasing numbers of endothelial cells toward the lumen, also indicating arterial dissection. We performed skin biopsy to investigate the skin collagen abnormalities. Electron microscopy revealed collagen bundles within the reticular dermis containing fibrils with highly variable, small diameters compared with normal control (Fig 2, C,D). Diseases causing arterial dissection were not evident. He was transferred elsewhere for rehabilitation on hospital day 65.

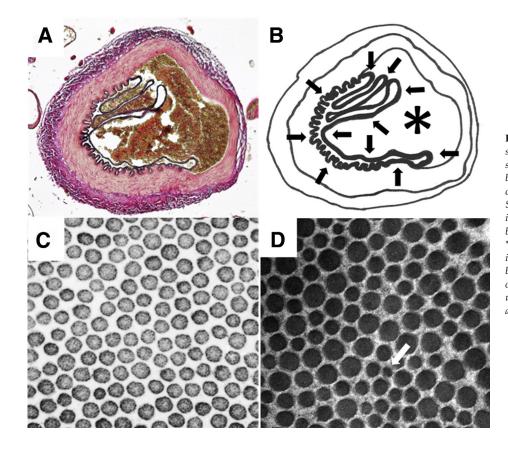


Figure 2. Elastica van Gieson (EVG) stain of middle cerebral artery M3 segment and appearance of collagen bundles. EVG staining of middle cerebral artery and M3 segment (A). Schema of artery (B) shows no inflammatory-cell invasion. Inner membrane has moved to left (black arrow). \*Red blood cells have accumulated below internal elastic lamina. Normal collagen bundles within reticular dermis consist of densely packed collagen fibrils with uniform cross sections (C), and white arrow shows irregular size of fibrils (D).

### Discussion

The unique point about the situation with this patient is that dissection was diagnosed from both a biopsy specimen of the MCA and from ultrastructural abnormalities of dermal connective tissue, whereas most MCAD has been diagnosed by angiography or at autopsy.<sup>1-4</sup>

To the best of our knowledge, this is the first description of finding dermal connective abnormalities in a skin biopsy of MCAD. Our findings, such as small diameter fibrils, were in line with those of prior reports in cervicocerebral artery dissection cases having collagen bundles within the reticular dermis containing several fibrils with irregular contours and morphometry on skin biopsies.<sup>6-10</sup> Most patients with cervicocerebral artery dissection accompanied by connective tissue disorders have subtle clinical manifestations such as hyperextensible skin or capillary fragility. Thus, skin biopsies might play an important role in confirming a diagnosis of cervicocerebral artery dissection.<sup>9</sup>

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