## Case Studies

## Adult-Onset Hemorrhagic Quasi-Moyamoya Disease with Unilateral Steno-occlusive Lesion in a Patient with Neurofibromatosis Type 1

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Background: Quasi-moyamoya disease is a condition that occurs in association with a specific underlying condition or disease such as atherosclerotic disease or neurofibromatosis type 1 (NF1). Pediatric cases are frequently reported, and an ischemic and bilateral presentation is more common than a hemorrhagic and unilateral presentation. Clinical Presentation: A 39-year-old woman previously diagnosed with NF1 presented to our department with nausea and left hemiparesis. She was diagnosed with right temporal intracerebral hemorrhage by initial computed tomography. Subsequent angiography showed an occlusion of the terminal portion of the right internal carotid artery, and magnetic resonance imaging showed multiple flow voids in the right basal ganglia, suggesting quasi-moyamoya disease. The hematoma was surgically removed, and her neurological condition improved after the operation. Conclusions: This is the first reported case of quasimoyamoya disease with a rare combination of characteristics, including an adult-onset, hemorrhagic presentation and a unilateral lesion in a patient previously diagnosed with NF1. Key Words: Carotid artery occlusion—hemorrhagic stroke—neurofibromatosis type 1—quasi-moyamoya disease. © 2018 National Stroke Association. Published by Elsevier Inc. All rights reserved.

Quasi-moyamoya disease is a relatively rare type of moyamoya disease that occurs in association with a specific underlying condition or disease such as atherosclerotic disease, autoimmune disease, or neurofibromatosis type 1 (NF1). Moyamoya disease is generally characterized by factors such as an adult or pediatric onset, an ischemic

or hemorrhagic presentation, and unilateral or bilateral lesions.

A 39-year-old woman previously diagnosed with NF1 presented to our emergency department with nausea and left hemiparesis. She was diagnosed with right temporal intracerebral hemorrhage by initial computed tomography (Fig 1, A,B). After admission to our department of neurological surgery, 3-dimensional computed tomography angiography on day 1 (Fig 1, C) and digital subtraction angiography on day 2 (Fig 1, D) showed an occlusion of the terminal portion of the right internal carotid artery (ICA). The hematoma was surgically removed on day 2, and the patient's hemiparesis improved as indicated by a change in her manual muscle testing grade from 1 to 2. Follow-up magnetic resonance imaging showed an occlusion of the C1-C2 portion<sup>2</sup> of the right ICA (Fig 1, E-H) and more than 2 flow voids in the right

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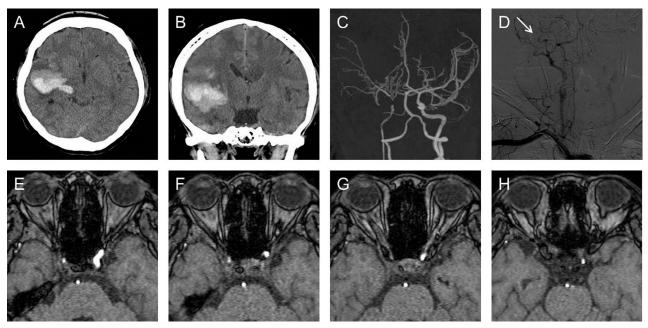
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**Figure 1.** Computed tomography images showing right temporal intracerebral hemorrhage (A, axial; B, coronal). (C) Three-dimensional computed tomography angiography and (D) digital subtraction angiography images showing an occlusion of the terminal portion of the right internal carotid artery. (E) C4, (F) C3, (G) C2, and (H) C1 portions of the internal carotid arteries on magnetic resonance angiography.

basal ganglia, suggesting unilateral quasi-moyamoya disease.<sup>1</sup> The patient was eventually discharged on day 22 without deterioration of her neurological condition or rebleeding.

Our patient had an occluded lesion in the terminal portion of the right intracranial ICA and more than 2 flow voids in the basal ganglia on magnetic resonance imaging, meeting the radiographic criteria for moyamoya disease.1 Quasi-moyamoya disease includes either a bilateral or a unilateral steno-occlusive lesion in the terminal portion of the ICA and is associated with underlying diseases such as NF1.1 Our patient had NF1, as well as specific unilateral radiographic characteristics that led to the diagnosis of unilateral quasi-moyamoya disease. Underlying conditions associated with quasi-moyamoya disease include atherosclerosis (29.0%), Down syndrome (15.1%), and NF1 (14.0%).1 Previous reports have described pediatric patients with NF1-associated moyamoya disease<sup>3-5</sup>; only a few reports have described adult cases.<sup>6,7</sup> In patients with NF1 who develop stroke, an ischemic presentation is more common than a hemorrhagic presentation.8 Additionally, bilateral lesions are much more common than unilateral lesions in patients with moyamoya disease.9 We have herein presented the first reported case of quasi-moyamoya disease with a rare combination of characteristics (adult-onset, hemorrhagic presentation and unilateral lesion) in a patient previously diagnosed with NF1.

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