Babinski-Nageotte Syndrome due to Vertebral Artery Dissection

Fumi Irie, Kazunori Toyoda, Noriko Hagiwara, Shigeru Fujimoto and Yasushi Okada

Abstract

Hemimedullary infarction, which presents Babinski-Nageotte syndrome, has been mainly reported to result from atherosclerotic occlusion of the vertebral artery. A 54-year-old housewife with right nuchal pain developed Wallenberg's syndrome followed by left hemiparesis. Diffusion-weighted magnetic resonance imaging documented fresh infarcts in the right hemimedulla and right dorsal cerebellum. Angiography revealed dissection of the right vertebral artery as a cause of the infarcts. Anterograde progression of the dissection might cause stepwise evolution of her neurological symptoms.

Key words: brain infarction, medulla oblongata, brainstem, Babinski-Nageotte syndrome, dissection, vertebral artery

Introduction

Since Babinski and Nageotte described an autopsy case of ischemic lesion involving the unilateral medulla oblongata one hundred years ago (1), the identity of hemimedullary stroke has been in dispute because both medial and lateral medullary infarctions rarely occur simultaneously and vascular supplies of medial and lateral medulla usually differ. The medulla oblongata can be divided into 4 zones according to its arterial circulation; the median zone which is perfused by the anterior spinal artery (ASA) at the level of lower medulla and the upper bulbar branches at the level of upper medulla, the paramedian zone by ASA, posterior inferior cerebellar artery (PICA) and middle bulbar branches, the lateral zone by PICA and lower bulbar branches, and the dorsal zone by PICA and posterior spinal artery (2, 3). Dissection of the vertebral artery (VA) is a possible pathology that involves ASA, PICA, and multiple bulbar branches and causes infarctions in both the medial and lateral medulla, although such infarctions due to dissection of the VA has not yet been reported in detail. Here we describe a patient with hemimedullary syndrome caused by unilateral VA dissection. Diffusion-weighted magnetic resonance imaging (DWI-MRI) was available for demonstration of her hyperacute ischemic lesion.

Case Report

A 54-year-old housewife without risk factors for arteriosclerosis including hypertension, diabetes, and hyperlipidemia started to suffer from right nuchal pain after one hour of swimming, which she habitually enjoyed three times a week. One month later, she suddenly felt nausea and then lost consciousness for several minutes at her home, and was admitted to a local hospital. She complained of vertigo then. The next morning, she suddenly developed weakness of the left limbs and was transferred to our stroke center.

On arrival 3 hours after progression of the symptoms, she was somnolent but cooperative and oriented. Her blood pressure was 184/88 mmHg and pulse rate was 66/min and regular. Her pupils were round and isocoric. Her ocular movements were full and smooth, with multidirectional nystagmus on vertical and horizontal gaze. There was right side paresis of her face, palate, and tongue. She was dysarthric and dysphagic, with impaired gag reflex. She had an almost complete motor palsy of the left limbs with increased left deep tendon reflexes and positive left Babinski reflex. Her right limbs were ataxic. Perception of pain and temperature was decreased on the right side of the face and left limbs. Deep sensations were intact. Paresthesia was present in all extremities.

DWI on the hospitalization day demonstrated hyperintense lesions at the right half of the medulla oblongata and the ipsilateral dorsal cerebellar hemisphere corresponding to PICA territory (Fig. 1A). Magnetic resonance angiography
Irie et al

Figure 1. A: Diffusion-weighted MRI on the hospitalization day. Areas of hyperintense signal in the right medial (arrow) and lateral (arrow) half of the medulla oblongata and ipsilateral dorsal cerebellar hemisphere (PICA territory) are demonstrated. B: Digital subtraction angiography of right VA on day 3. A frontal view shows a long segment of high-grade stenosis extending from C1 level with irregularity and occlusion at the base of the skull (arrow). C: T1-weighted (fat-suppressed) magnetic resonance imaging on day 9 of axial section demonstrates “crescent sign” in the right VA (arrow). D: MR angiography on day 22 revealed right VA and PICA (arrow), although indistinct compared with left VA.

(MRA) did not delineate right VA or right PICA. Cervical echography documented narrow right VA without diastolic flow signal, indicating occlusion of right VA proximal to right PICA (4). These neuroradiological studies did not document any parenchymal or vascular lesions in the anterior circulation. Blood tests were normal. Neither echocardiography nor prolonged electrocardiography revealed any cardiac source of emboli. Under probable diagnosis of dissection of right VA, we started continuous intravenous infusion of heparin (10,000 U/day, 14 days) within an hour after her arrival. The left hemiplegia suddenly recovered 2 hours later, but returned to the severe state again during the next 4 hours. Intravenous administration of urokinase (12,000 U) was then added. On day 3, right Horner’s sign became apparent.

On digital subtraction angiogram (DSA) on day 3, right VA became narrow from the C1 level and occluded at the level of foramen magnum (Fig. 1B). The basilar artery and its branches were intact via left VA. Anterior circulation was also intact. T1-weighted fat-suppressed MRI on day 9 showed a crescentic hyperintense area in the right VA at the level of foramen magnum indicating intramural hematoma in the false lumen (Fig. 1C). MRA on day 22 documented recanalization of right VA to the union (Fig. 1D). T2-weighted MRI on the same day delineated medullary and cerebellar infarcts in an area identical to that in the initial DWI. Based on these findings, we finally diagnosed right VA pathology as arterial dissection.

Her nystagmus and right facial paresis disappeared within 2 weeks. Bulbar palsy and motor and sensory deficits in the left limbs were improved to some extent. Ataxia in the right limb persisted. On discharge 3 weeks after admission, she needed a wheelchair for transfer.
**Discussion**

This is the first detailed case report of hemimedullary infarction due to VA dissection with radiological confirmation. Sudden onset of symptoms, tapering occlusion and later recanalization of VA, crescent sign on MRI, and lack of atherosclerosis or cardioembolic sources indicate VA dissection as a cause of infarction in this patient (5). Neck pain was reported to precede other manifestations of VA dissection for ≈ 12 days up to a maximum of 30 days, although headache preceded for a much shorter time (6). Thus in this case, the one-month history of nuchal pain after swimming might have been a warning sign of severe VA dissection.

Hemimedullary infarction is a rare disease. Twelve patients with infarction have been reported, to our knowledge, with pathological or radiological documentation (Table 1) (1, 3, 7–16). All patients had a unilateral medullary lesion and a patient by Nakane et al (13) had ipsilateral cerebellar infarct in the PICA territory, like the present patient. VA occlusion was a leading vascular pathology, caused by syphilis and endarteritis in earlier reports, and by atherosclerosis with hypertension or diabetes in recent reports. VA dissection was reported as an etiology of hemimedullary infarction in only one study, without a detailed case presentation (3).

Spontaneous VA dissection occurs in 1 to 1.5 per 100,000 person-years, and is an important cause of ischemic stroke in young and middle-aged patients (5). This vascular disease is a frequent cause of lateral medullary infarction. A neuro-radiological study indicates that more than half of patients with lateral medullary infarction had definite or probable VA dissection (17). Contrarily, the dissection was rarely reported to cause medial medullary infarction (18, 19). In our analysis of 11 patients with medial medullary infarction, only one patient (9%) was suspected to have VA dissection (19). Another study however, reported the 3 among 7 patients (43%) with medial medullary infarction had spontaneous extracranial VA dissection (3). Thus, the frequency of VA dissection in medial medullary infarction has been disputed. Because bilateral ASAs usually supply the medial medulla and unilateral PICA supplies the lateral medulla, unilateral VA dissection does not seem to cause medial medullary infarction as often as lateral one. The initial symptoms for the present patient correspond to PICA-territorial syndrome. Pyramidal symptoms, resulting from medial medullary infarction, appeared on the second day. The time course suggests anterograde progression of her arterial dissection; it might have reached the orifice of PICA on the day of stroke onset, and the orifice of ASA and upper bulbar branches on day 2.

In early clinical reports, hemimedullary syndrome was generally fatal, especially if complicated with respiratory disturbances. The syndrome was so uncommon that it was difficult to make the topographical and etiological diagnoses while patients were alive. Here, the advent of new neuroimaging strategies including DWI and MRA led to definite topographical diagnosis and the probable etiological diagnosis as arterial dissection and to the immediate start of treatment using urokinase and heparin within 4 hours after stroke progression of this patient. The quick treatment might cause transient recovery from the hemiparesis, but did not end in good functional outcome without assistance on discharge. Although intravenous and local intra-arterial thrombolysis is feasible in stroke caused by cervical arterial dissection (20, 21), further studies are necessary to establish

<table>
<thead>
<tr>
<th>Authors</th>
<th>Year</th>
<th>Site of infarct</th>
<th>Site of arterial occlusion</th>
<th>Etiology</th>
<th>Means of diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Babinski and Nageotte [1]</td>
<td>1902</td>
<td>L</td>
<td>LVA (distal end), BA</td>
<td>Meningovascular syphilis</td>
<td>Autopsy</td>
</tr>
<tr>
<td>Marinesco and Draganesco [8]</td>
<td>1923</td>
<td>R</td>
<td>RVA, stenosis of BA</td>
<td>Atherosclerosis, Lipoid, crystalline</td>
<td>Autopsy</td>
</tr>
<tr>
<td>Pines and Gilinsky [9]</td>
<td>1930</td>
<td>L</td>
<td>LVA</td>
<td>Thrombus</td>
<td>Autopsy</td>
</tr>
<tr>
<td>Harris and Hauser [10]</td>
<td>1931</td>
<td>R</td>
<td>stenosis of RVA (distal end)</td>
<td>Thrombus, infection</td>
<td>Autopsy</td>
</tr>
<tr>
<td>De Freitas et al [16]</td>
<td>2001</td>
<td>R</td>
<td>Severe stenosis of RVA</td>
<td>Hypertension, diabetes</td>
<td>MRI, MRA</td>
</tr>
</tbody>
</table>

R: right, L: left, VA: vertebral artery, BA: basilar artery.
the appropriate acute management.

Acknowledgements: This study was partially supported by the Japanese Ministry of Health, Labour and Welfare (12C-2).

References