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CASE REPORT

Rheumatoid Arthritis–Induced Lateral Atlantoaxial Subluxation With Multiple Vertebrobasilar Infarctions

Yasuhiro Takeshima, MD, PhD, Ryosuke Matsuda, MD, PhD, Yasuo Hironaka, MD, PhD, Yasushi Motoyama, MD, PhD, and Hiroyuki Nakase, MD, PhD

Study Design. Case report.

Objective. To highlight the probability that lateral atlantoaxial subluxation (AAS) exists in patients with rheumatoid arthritis (RA) and induces vertebrobasilar infarctions that are more foregrounded than compressive myelopathy.

Summary of Background Data. Although lateral subluxation is a well-known subtype of AAS, a case of cerebral ischemia associated with lateral AAS has not been reported before.

Methods. A 52-year-old male with a 6-year history of RA had a sudden onset of visual field defect and mild right cerebellar ataxia. Head magnetic resonance imaging revealed acute multiple infarctions in the vertebrobasilar area, and magnetic resonance angiography revealed stenosis of the left vertebral artery (VA). Lateral radiograph of the cervical spine in the neutral position revealed atlanto-occipital assimilation and anterior AAS. T2-weighted sagittal images on cervical magnetic resonance imaging revealed high signal intensity in the spinal cord at C1–C2. Cerebral angiography revealed right VA occlusion and severe stenosis of the left V3 segment of VA. Three-dimensional computed tomography angiography of the craniovertebral junction revealed lateral AAS, which was due to severe erosive changes of the facet joints, and the left V3 portion was stenosed by a bony component. During conservative therapy, the patient experienced left occulomotor nerve palsy due to a second stroke.

Results. Two months later, the patient underwent occipitocervical posterior fusion with an iliac bone graft. His postoperative course was uneventful, and the left VA stenosis disappeared. At the 45-month follow-up, he had no further infarctions. Bony fusion was radiologically confirmed, and 3-dimensional computed tomography angiography revealed good patency of the affected left VA.

Conclusion. In patients with RA, the potential risk of AAS should be recognized. Lateral AAS in particular may induce cerebral ischemia by positional VA occlusion in advanced stages of the disease.

Key words: rheumatoid arthritis, multiple cerebral infarction, vertebral artery, atlantoaxial subluxation, positional vertebral artery occlusion, 3-dimensional computed tomography angiography, lateral subluxation, posterior fusion.

Level of Evidence: N/A

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Atlantoaxial subluxation (AAS) with severe occipitalgia or high cervical myelopathy is the most common type of instability of upper cervical involvement in patients with rheumatoid arthritis (RA).[1] A subtype of this disorder is lateral subluxation, which usually occurs with rotational deformity and is observed in 20% of cases.[2,3] However, to the best of our knowledge, there are no reported cases of cerebrovascular disease induced by lateral AAS in patients with RA. Here, we present a unique case of multiple vertebrobasilar infarctions associated with RA-induced lateral AAS that are more prominent than compressive myelopathy.

CASE REPORT

A 52-year-old male presented with sudden visual disturbance. He had a 6-year history of RA with severe osteolytic changes in his knee and hand joints with disability, classified into Steinbrocker stage 3,[4] a longtime steroid use, and a history of cerebral infarctions with aftereffects of right hemiparesis. Neurological examination revealed left hemianopsia and mild right cerebellar ataxia. His head was in a position similar to "cock robin" deformity, with intense dizziness and nausea during side-to-side head movement. Deep tendon reflexes remained normal, but he experienced glove-and-stocking–type numbness. Head magnetic resonance imaging (MRI)

From the Department of Neurosurgery, Nara Medical University School of Medicine, Kashihara, Japan.

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Address correspondence and reprint requests to Yasuhiro Takeshima, MD, PhD, Department of Neurosurgery, Nara Medical University School of Medicine, 840 Shijo-cho, Kashihara, Nara, 634-8521, Japan; E-mail: takeshim@naramed-u.ac.jp

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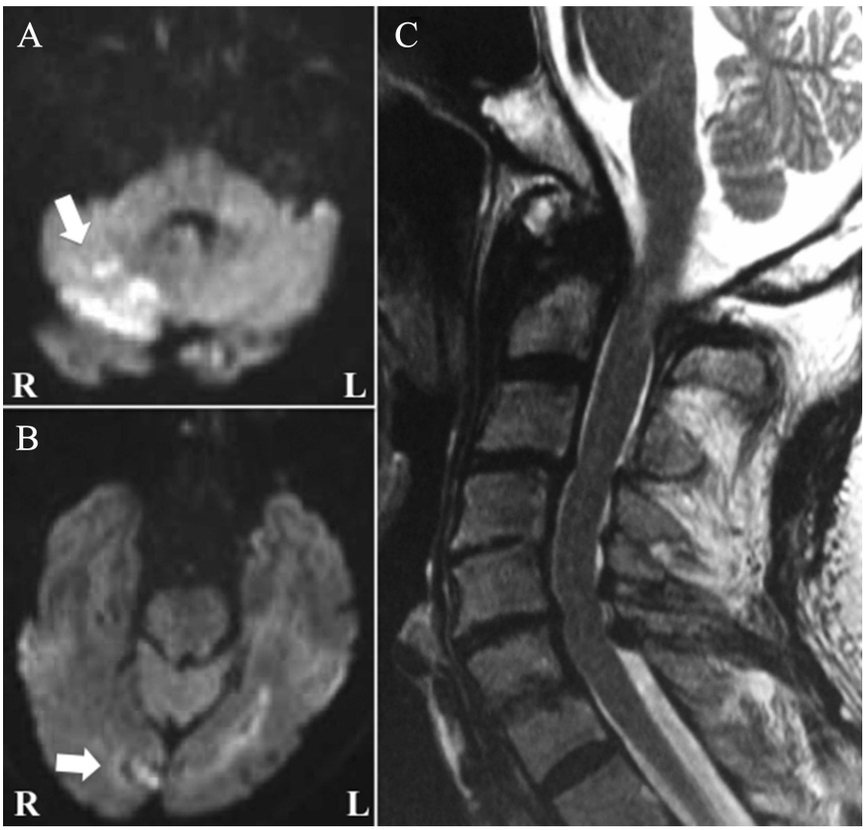

Figure 1. Magnetic resonance images of the brain and cervical spine at onset. **A**, **B**, High-intensity lesions in the cerebellum and right occipital lobe on an axial diffusion-weighted image revealing multiple acute infarcts (arrows). **C**, Destruction of the dens of axis and intramedullary hyperintensity in the upper cervical cord on the sagittal T2-weighted image.

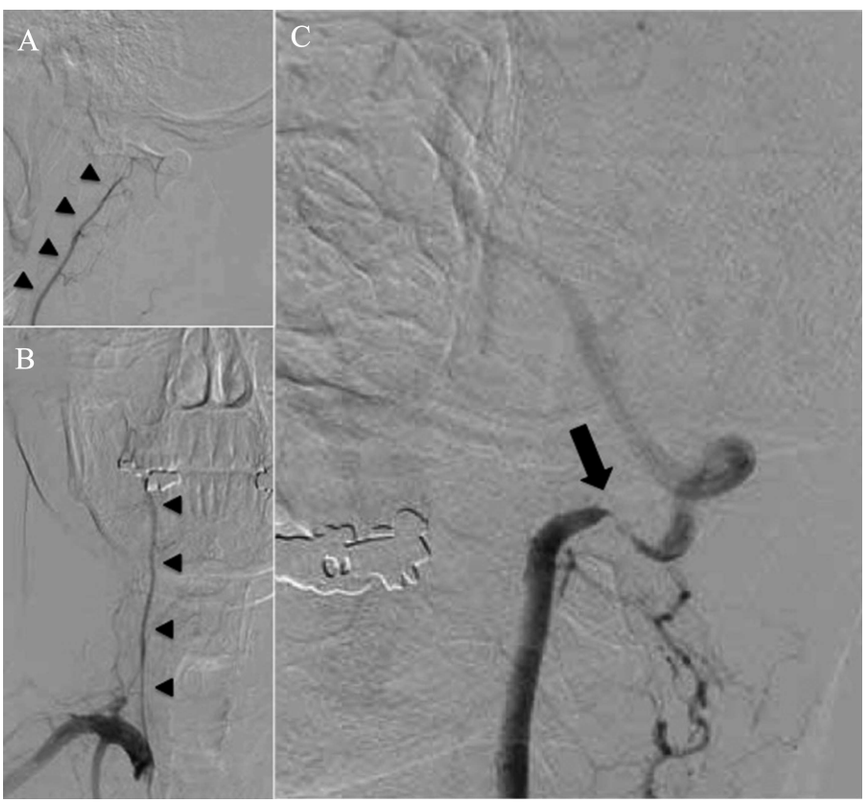

Figure 2. Preoperative VAG. Lateral (**A**) and anteroposterior (**B**) views on right VAG reveal hypoplastic right vertebral artery (arrowheads). Oblique view on left VAG (**C**) reveals severe stenosis of the left V3 portion (arrow). VAG indicates vertebral angiography.

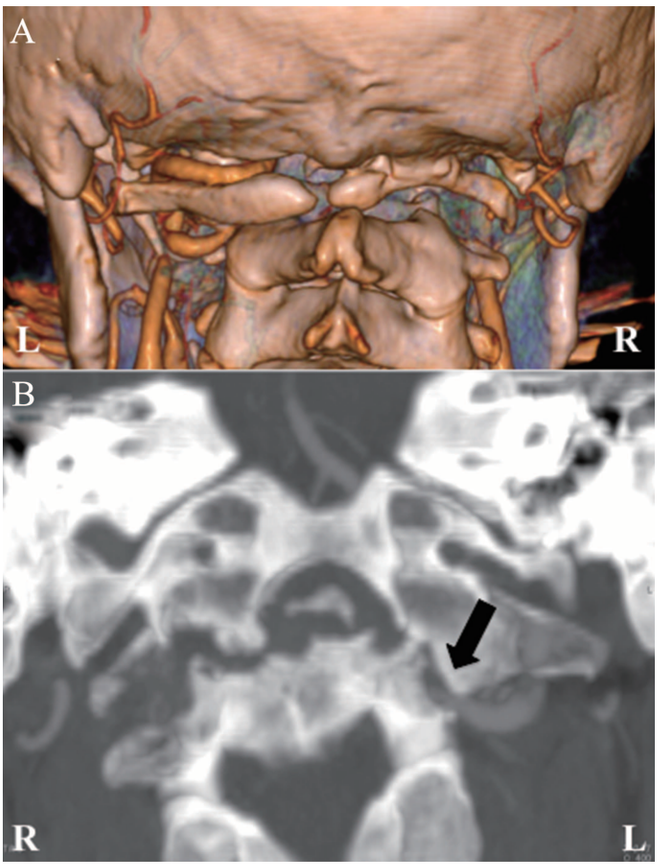

Figure 3. 3D-CTA of craniovertebral junction. **A**, Posterior 3D-CTA view reveals lateral subluxation of C1–C2 and dysraphism of the C1 posterior arch. **A**, Coronal reconstruction image of the craniovertebral joint reveals erosive change in the lateral subluxation of C1–C2; the left vertebral artery was osseously wedged at the transverse foramen of the axis (arrow). 3D-CTA indicates 3-dimensional computed tomography angiography.

revealed multiple acute infarctions in the right cerebellar hemisphere and occipital lobe (Figure 1A, B), and magnetic resonance angiography revealed stenosis of the extracranial portion of the left vertebral artery (VA). Lateral radiograph of the cervical spine in the neutral position showed occipitalization of the atlas and an atlantodental interval of 10 mm. Further MRI scans of the cervical spine revealed intramedullary hyperintensity on T2-weighted images at the atlantoaxial level (Figure 1C). Occlusion of the right VA (Figure 2A, B) and severe stenosis of the third segment (V3) of the left VA (Figure 2C) were confirmed by cerebral angiography. Coronal view reconstruction of 3-dimensional computed tomography angiography at the craniovertebral junction revealed lateral subluxation and osseous compression of the left VA, which was due to severe erosive change of the atlantoaxial facet joints (Figure 3A, B).

We performed heparinization and external fixation with a hard neck collar during these examinations; however, he had a new pontine infarction with left oculomotor nerve palsy without a trigger event. Two months later, occipitocervical posterior fusion with an iliac bone graft was performed (Figure 4A). To prevent further mechanical injury to VA, the stenotic portion of the artery was not exposed during surgery and the blood flow of the left VA was monitored with a Dopplar flowmeter. Heparinization was reinitiated the day after the operation, and anticoagulant therapy with warfarin was continued. The postoperative course was uneventful, and he had no recurrent cerebrovascular stroke at the 45-month follow-up. Bony fusion was radiologically confirmed, and 3-dimensional computed tomography angiography revealed disappearance of the stenosis of the left VA (Figure 4B).

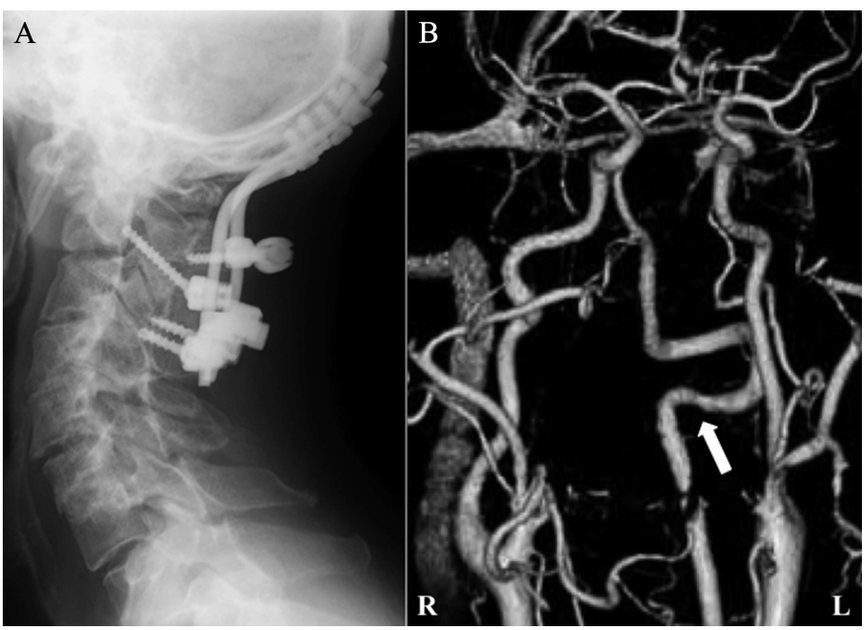

Figure 4. Postoperative lateral radiograph of the cervical spine (**A**) and an oblique view of 3-dimensional computed tomography angiography (**B**). Computed tomography angiography reveals complete disappearance of the stenosis of the left V3 portion (arrow) compared with preoperative left vertebroarteriography (Figure 2C).

DISCUSSION

In patients with RA, the upper cervical spine is the most common site of damage, causing severe neck pain, myelopathy, and, in some cases, sudden death.[5] Webb *et al*[6] were the first to report AAS-induced fatal thromboembolic events in patients, which was discovered at autopsy. Anterior subluxation is the most common type of AAS; however, posterior and lateral subluxation subtypes also occur occasionally. Although there are few case reports on AAS-induced vertebrobasilar insufficiency in patients with RA,[1,6–8] all these are not of the lateral type. Abnormal rotation of the atlantoaxial joint often accompanies lateral subluxation; however, the conception that lateral AAS may injure VA did not exist until now. Its assessment is often difficult because, occasionally, optimal frontal visualization of atlantoaxial facet joints is difficult[3]; therefore, the possible existence of lateral AAS with vertebrobasilar insufficiency in patients with RA should be considered.

Consideration for the mechanism of cerebral embolism in the present case is quite interesting. The underlying pathology of the case was positional VA occlusion, a condition classically characterized by temporary hemodynamic cerebral ischemia.[9] On the contrary, previous studies have recently reported cases of embolic ischemia inducing artery-to-artery cerebral embolisms after arterial intimal injury.[10–13] It was presumed that the chronic progression of RA gradually induced atlantoaxial arthritis and instability. Furthermore, mechanical compression of VA due to lateral subluxation of the atlantoaxial joint caused intimal injury and created embolic sources at that lesion, indicating cerebral artery-to-artery embolism. But no apparent embolus or intimal injury was detected by diagnostic imaging.

Myelopathy is a major reason for surgical intervention or indication for surgery, but stroke event has not been generally paid attention in patients with RA with AAS. Because severe physical impairment may occur after a cerebral stroke, the possibility of vertebrobasilar insufficiency should be considered with or without myelopathy in such patients.

➢ Key Points

- ❑ Lateral AAS presenting with cerebral stroke is a very rare clinical condition.
- ❑ Three-dimensional computed tomography angiography clearly revealed osseous compression of the left VA at the axial level.
- ❑ The potential risk of AAS should be recognized in patients with RA because the lateral form of this disorder may induce cerebral ischemia.

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