Case Studies

Progressive Bilateral Vertebral Artery Dissection in a Case of Osteogenesis Imperfecta

Yuji Kato, MD, PhD,* Harumitsu Nagoya, MD, PhD,* Tetsuya Abe, MD,* Takeshi Hayashi, MD, PhD,* Masanori Yasuda, MD, PhD,† Akira Uchino, MD, PhD,‡ Norio Tanahashi, MD, PhD,* and Masaki Takao, MD, PhD*

A 32-year-old woman with osteogenesis imperfecta (OI) was admitted to the hospital because of a right-sided occipital headache and facial paresthesia. She was diagnosed with lateral medullary syndrome due to right vertebral artery (VA) dissection. She was treated conservatively without antithrombotic therapy. She developed subarachnoid hemorrhage because of contralateral VA dissection 18 days later. This clinical course may reflect the underlying weakness of the vessel wall in OI. In patients with OI, occlusion of a unilateral VA could cause dissection and subsequent rupture of the contralateral VA. Early surgical treatment for lesions of the VA is required in such cases. **Key Words:** Osteogenesis imperfecta—bilateral vertebral artery dissection—lateral medullary syndrome—subarachnoid hemorrhage.

Introduction

Osteogenesis imperfecta (OI) is a heterogeneous and inherited connective tissue disorder that is characterized by excessive bone fragility.[1] OI is caused by abnormalities of type I collagen, which is a major structural component of vessel walls. Nevertheless, cerebrovascular complications have not been well analyzed in OI.[2-7] We present a case of progressive bilateral vertebral artery (VA) dissection in a patient with OI.

Case Report

A 32-year-old woman developed a right-sided occipital headache and facial paresthesia. The patient had clinically diagnosed OI (type 1) because of the presence of bone fragility, blue-tinged sclerae, a family history, and bone X-ray findings, including Wormian bone and low bone density.[1,8] She had a medical history of fractures of the limbs (4 times), which were associated with minor trauma since she was 4 years old. Her father and elder brother also had a history of multiple fractures and were clinically diagnosed with OI. The family history of the patient suggested autosomal dominant transmission (Fig 1). She had no risk factors, such as hypertension, dyslipidemia, arrhythmia, or other cardiovascular diseases, or diabetes mellitus.

She was alert on admission and her height was 140 cm. Her blood pressure was 127/76 mmHg and her heart rate was 84 beats/minute. A neurological examination showed right-sided blepharophimosis, miosis, and facial anhidrosis.

From the *Department of Neurology and Cerebrovascular Medicine, Saitama Medical University International Medical Center, Hidaka, Saitama, Japan; †Department of Pathology, Saitama Medical University International Medical Center, Hidaka, Saitama, Japan; and ‡Department of Diagnostic Radiology, Saitama Medical University International Medical Center, Hidaka, Saitama, Japan.

Received October 29, 2016; revision received December 2, 2016; accepted December 17, 2016.

Address correspondence to Yuji Kato, MD, PhD, Department of Neurology and Cerebrovascular Medicine, Saitama Medical University International Medical Center, 1397-1 Yamane, Hidaka, Saitama 350-1298, Japan. E-mail: yujik@saitama-med.ac.jp.

1052-3057/$ - see front matter

http://dx.doi.org/10.1016/j.jstrokecerebrovasdis.2016.12.012

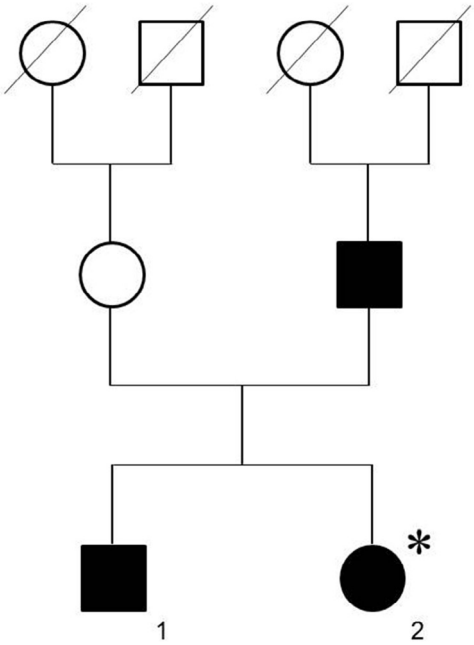

Figure 1. *Abbreviated family pedigree. The square indicates a man and the circle indicates a woman. Dark squares and circles indicate a history of multiple fractures. An asterisk indicates the present case.*

Rotatory nystagmus in the fast phase was directed toward the left side. She had dysphagia resulting from soft palatal paresis. The gag reflex had disappeared on the right side, as well as dysmetria on the right side by finger-to-nose testing, and ataxic gait.

Diffusion-weighted magnetic resonance (MR) imaging on admission showed acute infarction at the right posterolateral medulla (Fig 2, A). T1-weighted MR imaging, MR angiography, and partial maximum intensity projection (MIP) images of the vertebrobasilar system showed thrombotic occlusion in the right VA and dilation of the proximal basilar artery (BA) (Fig 2, B-D). An MR angiographic source image showed a double lumen in the occluded right VA (Fig 2, E). Basi-parallel anatomical scanning-MR imaging[9] showed dilatation of the outer contour at the right VA and proximal BA (Fig 2, F). Three-dimensional computed tomography angiography on admission showed almost complete occlusion in the right VA and dilation of the proximal BA (Fig 3, A), although there appeared to be retrograde residual flow (Fig 3, B). Based on these findings, we diagnosed lateral medullary syndrome due

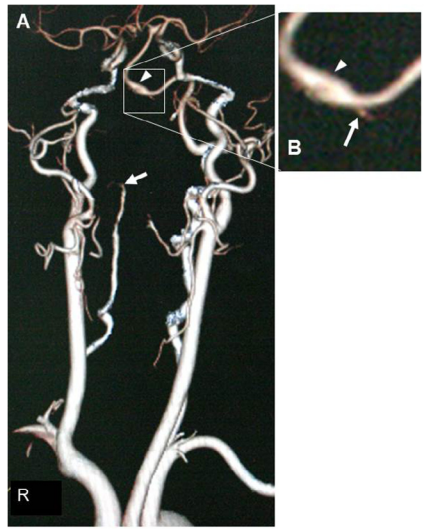

Figure 3. *Three-dimensional computed tomography angiography on admission shows occlusion in the right vertebral artery (A, **arrow**) and dilation of the proximal basilar artery (A, B, **arrowhead**), although there appears to be retrograde residual flow (B, **arrow**).*

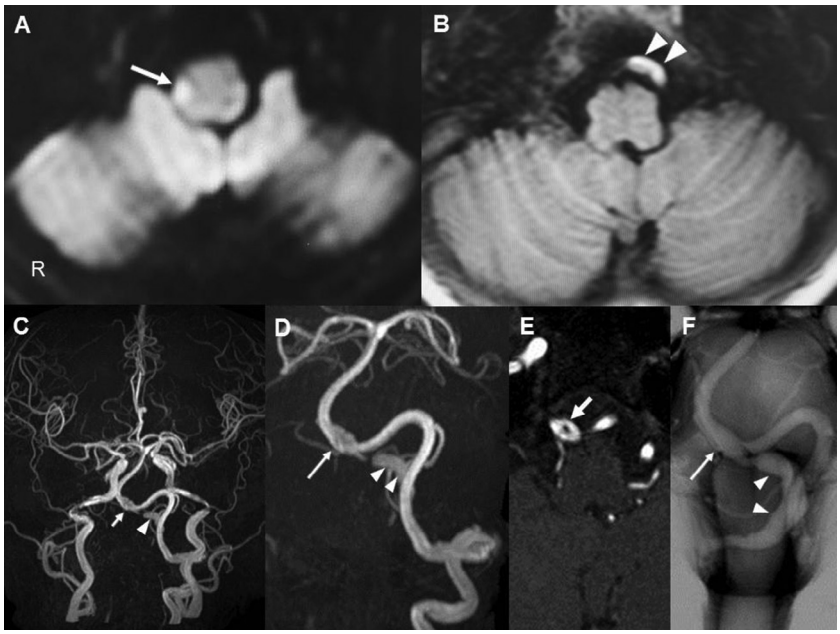

Figure 2. *Diffusion-weighted MR imaging (A) performed on admission shows hyperintensity in the right posterolateral medulla (**arrow**). T1-weighted MR imaging (B), MR angiography (C), and a partial maximum intensity projection image of the vertebrobasilar system (D) show occlusion of the right VA with thrombus in the pseudolumen (**arrowheads**) and dilation of the proximal basilar artery (BA) (**arrow**). MR angiographic source image (E) shows a double lumen in the right occluded VA (**arrow**). Basi-parallel anatomical scanning-MR imaging (F) shows dilatation of the outer contour at the right VA (**arrowheads**) and the proximal BA (**arrow**). Abbreviations: MR, magnetic resonance; VA, vertebral artery.*