



A Juvenile Case of Bow Hunter's Syndrome Caused by Atlantoaxial Dislocation with Vertebral Artery Dissecting Aneurysm

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Key words

- Atlantoaxial rotatory dislocation
- Bow hunter's syndrome
- C1-C2 posterior fusion
- Dissecting aneurysm
- Vertebral artery

Abbreviations and Acronyms

AAD: Atlantoaxial dislocation
BHS: Bow hunter's syndrome
SAH: Subarachnoid hemorrhage
VA: Vertebral artery

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INTRODUCTION

Bow hunter's syndrome (BHS) is a rare symptomatic vertebrobasilar insufficiency that occurs during neck rotation¹ and is often caused by a bony abnormality that compresses the vertebral artery (VA), compromising distal flow, or leads to a vessel wall injury resulting in thromboembolism. It is reported that BHS is often caused by atlantoaxial dislocation (AAD).² Resulting symptoms affect patients' activities of daily living owing to infarction in the cerebellum or brainstem. Management of BHS is dependent on each patient. First, a conservative approach, such as neck immobilization and antiplatelets or anticoagulation, is followed to prevent secondary stroke. When this approach is not effective, a surgical approach, such as C1-C2 posterior fusion or decompression of the VA, is planned.³ We report the case of a teenage girl who experienced an acute cerebral infarction in the right thalamus and a sudden occlusion of the

■ **BACKGROUND:** Bow hunter's syndrome (BHS) is caused by posterior circulation insufficiency that results from the occlusion or compression of the vertebral artery (VA) during neck rotation. Owing to its rarity, there is no guideline to support the decision of selecting a conservative or a surgical approach. Management of BHS is dependent on each patient.

■ **CASE DESCRIPTION:** A 13-year-old girl presented with transient visual disturbance, hypoesthesia, and paralysis of the left side of the body. Magnetic resonance imaging revealed an acute cerebral infarction in the right thalamus, and magnetic resonance angiography demonstrated occlusion of the right posterior cerebral artery and dilation of V3 of the left VA. Digital subtraction angiography revealed a left VA dissecting aneurysm at V3 and left VA occlusion at the level of C1-C2 during neck rotation to the right. A dynamic x-ray suggested atlantoaxial joint instability, and three-dimensional computed tomography revealed aplasia of C1 lamina and atlantoaxial rotatory dislocation. BHS with left VA dissecting aneurysm caused by atlantoaxial rotatory dislocation was diagnosed. We performed C1-C2 posterior fusion by the Goel-Harms technique. Stroke did not recur, and computed tomography angiography obtained 8 months postoperatively demonstrated a decrease in the dissecting aneurysm.

■ **CONCLUSIONS:** To our knowledge, this is the first case of BHS with VA dissecting aneurysm and aplasia of C1 lamina. Based on this case, we suggest that C1-C2 posterior fusion is effective for BHS with VA dissecting aneurysm.

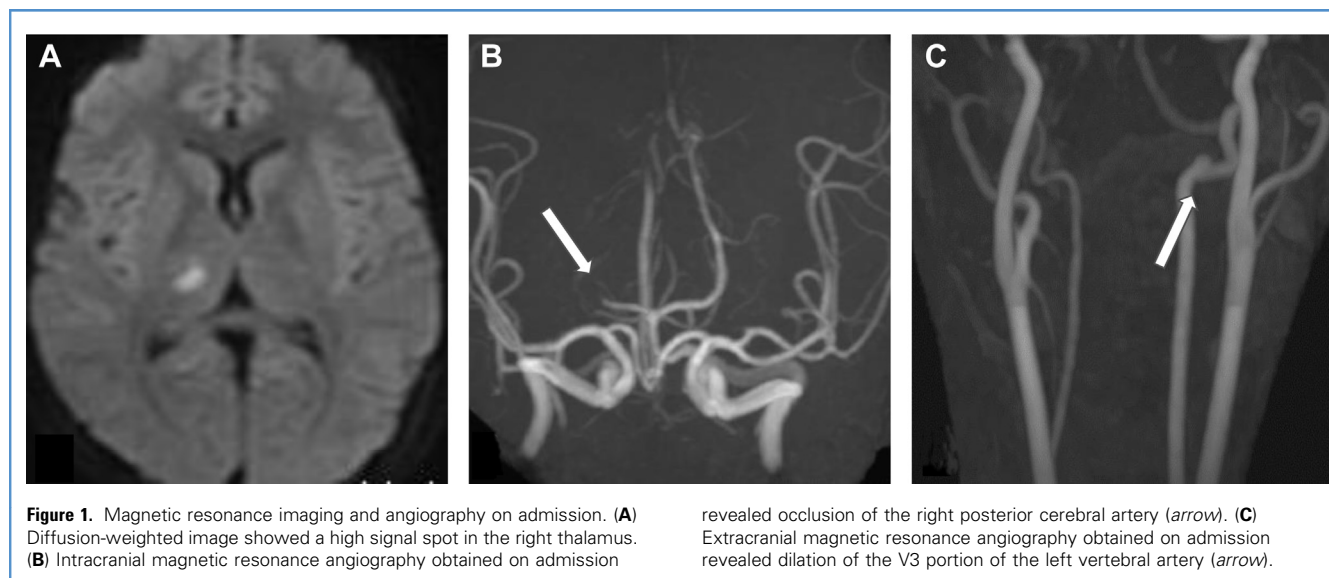
right posterior cerebral artery. We diagnosed BHS with dissecting aneurysm at the V3 portion of the left VA and rotatory AAD caused by aplasia of C1 lamina. The repetitive mechanical stimulation of the left VA was suspected to have caused BHS with a dissecting aneurysm. To prevent recurrent ischemic stroke, we performed C1-C2 posterior fusion using the Goel-Harms technique. A second stroke did not occur. To our knowledge, this is the first report of BHS with a VA dissecting aneurysm and aplasia of C1 lamina. Based on this case report, we suggest that C1-C2 posterior fusion is effective for BHS with VA dissecting aneurysm.

CASE DESCRIPTION

A 13-year-old girl presented with sudden and transient visual disturbance, hypoesthesia, and paralysis of the left side of

the body. She denied other symptoms and had no history of trauma. Physical examination revealed left mild hemiparesis and slight hypoesthesia of the left side of the body. Blood and coagulation tests (including platelet count, prothrombin time, international normalized ratio, activated partial thromboplastin time, fibrinogen, antithrombin III, D-dimer) showed no abnormalities. Electrocardiogram showed normal sinus rhythm. Magnetic resonance imaging and magnetic resonance angiography revealed acute cerebral infarction in the right thalamus and occlusion of the right posterior cerebral artery, respectively. Extracranial magnetic resonance angiography demonstrated dilation of the V3 portion of the left VA. We suspected this to be the lesion of the dissecting aneurysm (Figure 1).

The cause of ischemic infarction was considered to be artery-to-artery embolism from the dissecting aneurysm at the V3



portion of the left VA. Aspirin (100 mg/day) was initiated as a conservative approach to prevent secondary stroke. In the search for the embolic source, echocardiography demonstrated normal left ventricle function and no wall motion abnormality. Carotid sonography demonstrated the flow disruption of the left VA during neck rotation to the right, although neck rotation did not cause any ischemic symptoms. During digital subtraction angiography, right vertebral and bilateral cerebral angiography revealed no abnormality, whereas left vertebral angiography revealed no blood flow stasis and vessel dilation at the V₃ portion. Three-dimensional rotational angiography revealed a 6.5-mm aneurysm. During neck rotation to the right, left vertebral angiography showed that the blood was completely occluded at the level of C₁-C₂ (**Figure 2**). Cervical x-ray showed an asymmetry in the atlanto-odontoid distance. Dynamic x-ray in a lateral view demonstrated an atlantodental interval of 5.5 mm at the flexion position (i.e., atlantoaxial joint instability) and 3.3 mm at the neutral position (**Figure 3**). Three-dimensional computed tomography revealed aplasia of C₁ lamina and C₁-C₂ facet dislocation on the left side of the neutral position. This dislocation worsened during right head rotation (**Figure 4**).

Two months after the onset, we performed C₁-C₂ posterior fusion using the Goel-Harms technique with navigation

systems to prevent recurrent ischemic stroke and reduce the stimulation of the dissecting aneurysm and VA. We inserted screws on both sides of the C₁ lateral mass and C₂ pedicle. Then we inserted a bone graft from iliac bone on the posterior arch of C₁ to C₂; the graft was fixed using polyethylene sutures (**Figure 5**).

The patient was managed with neck immobilization for 3 months and antiplatelet therapy (aspirin 100 mg/day) to prevent ischemic stroke for 6 months. Her neurologic symptoms resolved, and she experienced only slight hypoesthesia of the left hand. Stroke did not recur, and follow-up computed tomography angiography performed over 8 months postoperatively revealed decreasing dissecting aneurysm and improving wall irregularity (**Figure 6**).

DISCUSSION

BHS is caused by posterior circulation insufficiency resulting from the occlusion or compression of VA during neck rotation.¹ The mechanism of ischemic stroke is not only hemodynamic but also thrombogenic owing to repetitive injury to the VA. This leads to thrombosis and dissection, aneurysm formation, and subsequent distal embolization.⁴

The etiology of BHS includes atlanto-occipital membrane tethering the VA, AAD, trauma, cervical disc herniation, rheumatoid arthritis, tumor, infection,

and Down syndrome.² In this study, the cause of BHS was considered to be AAD owing to aplasia of C₁ lamina, which is a congenital abnormality observed in 4% of autopsy samples.⁵ Aplasia of C₁ lamina may cause atlantoaxial instability and compromised spinal cord, which leads to neurologic symptoms. For this abnormality, a surgical approach is decided based on the presence of atlantoaxial instability or neurologic symptoms.⁶

First, a conservative approach such as neck immobilization and antiplatelets or anticoagulation is followed to prevent secondary stroke. Owing to the rarity of BHS, there is no guideline to support the decision of selecting a conservative or a surgical approach.

When a conservative approach is ineffective or affects the ability of the patient to perform activities of daily living, a surgical approach becomes necessary. Two techniques, C₁-C₂ posterior fusion and decompression of the VA, are available. Fujimoto et al.³ described these 2 techniques. They also reported that posterior fusion is more complicated than decompression. Posterior fusion reduces the range of head rotation. The head rotation is said to be 60° to each side, 120° in total, and 70% of this rotation is attributed to the C₁-C₂ joint. Fujimoto et al. performed C₁-C₂ posterior fusion in 12 patients. The rate of head rotation reduction was 50%–70% (average

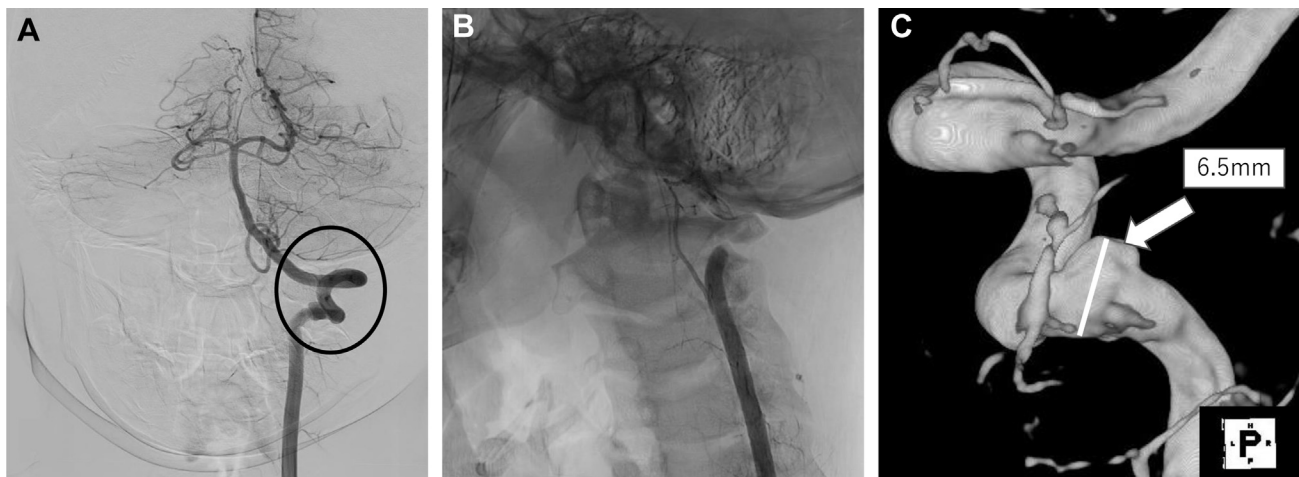


Figure 2. Digital subtraction angiography. (A) Left vertebral angiography in the neutral head position showed no stenosis-occlusive lesion and dilation of the V3 portion (circle). (B) Left vertebral angiography in the right head

rotation showed that blood flow was completely occluded at the level of C1-C2. (C) Three-dimensional rotational angiography revealed a 6.5-mm aneurysm (arrow). P indicates the posterior side of the patient.

58.8%). This rotation was reported to not inhibit patients' activities of daily living. In contrast, decompression does not limit head rotation. However, Fujimoto et al. reported that the patients had recurrence of stroke and needed C1-C2 posterior fusion because the decompression of the VA may have caused the VA to adhere to surrounding structures, leading to tethering of the VA. Regarding the surgical approach for BHS, they concluded that C1-C2 posterior fusion was more useful and safer than decompression.³

The symptoms of VA dissection are classified into 4 categories: 1) VA occlusion, 2) thromboembolism caused by clot from inner membrane damage or pseudolumen by dissection, 3) subarachnoid hemorrhage (SAH) by dissecting aneurysm rupture, and 4) dissecting aneurysm compressing the brainstem or nerve. Treatments for VA occlusion and thromboembolism include anticoagulants or antiplatelets for preventing the expansion of ischemic stroke and formation of clots. Treatments for SAH and dissecting

aneurysm include an endovascular or surgical approach.⁷

The histopathologic characterization of VA and the symptoms of VA dissection are different intracranially and extracranially. Intracranial VA has thin outer and medial membranes and lacks elastic fiber at the external elastic lamina and medial membrane.⁸ This characteristic structure causes VA dissection in the medial membrane alone or between outer and medial membranes. Intracranial VA dissection often causes SAH.⁹ Treatments for

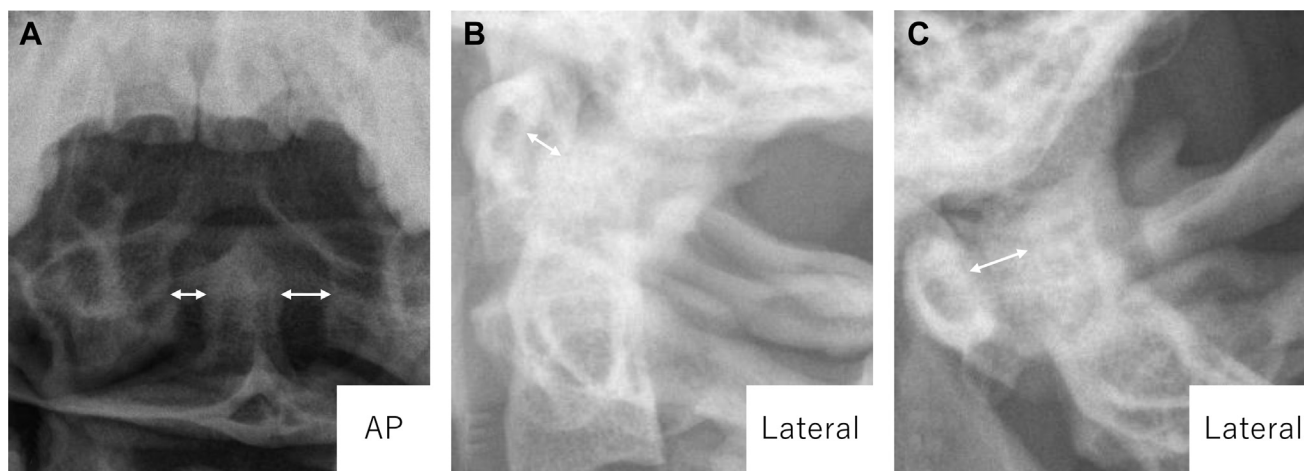


Figure 3. Cervical x-ray and dynamic x-ray images. (A) Cervical x-ray in an open-mouth position showed asymmetry in the atlanto-odontoid distance (arrows). (B) Cervical x-ray in a lateral view showed that the atlantodental

interval was 3.3 mm at the neutral position (arrow). (C) Cervical x-ray in a lateral view showed that the atlantodental interval was 5.5 mm at the flexion position (arrow).

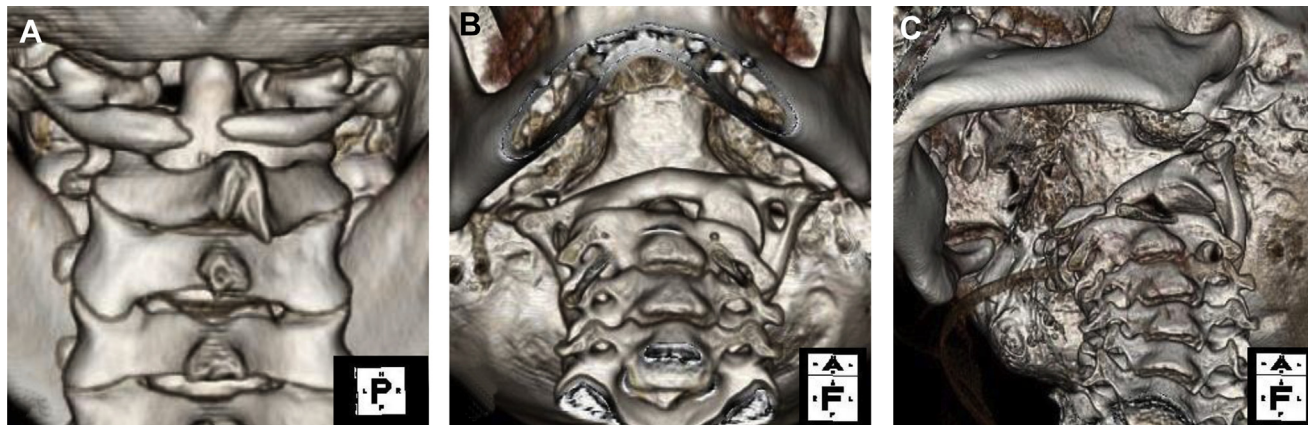


Figure 4. Three-dimensional computed tomography (3D-CT). (A) 3D-CT showed aplasia of C1 lamina. (B) 3D-CT showed atlantoaxial rotatory dislocation and C1-C2 dislocation on the left facet in the neutral position. (C)

3D-CT showed atlantoaxial rotatory dislocation worsened due to head rotation to the right. A and P indicate the anterior and posterior sides of the patient, respectively; F indicates toward the feet of the patient.

intracranial VA dissection include surgical and endovascular approaches to prevent SAH.^{10,11}

In contrast, extracranial VA has a thick external and internal elastic lamina, outer membrane with rich collagen fiber, medial membrane with rich elastic fiber, and developed muscular layer.⁶ Extracranial VA dissection leads to intramembrane hematoma and inner membrane damage, causing VA stenosis or thrombotic ischemic stroke. They often occur at the level of the atlantoaxial joint, which is likely to be affected by head rotation, and the foramen transversarium of the

sixth cervical vertebra, which is likely to be affected by neck extension.⁹ Treatment for extracranial VA dissection includes a conservative approach, such as antiplatelets or anticoagulants, first for secondary stroke prevention. When recurrence of ischemic stroke or worsening of VA dissection is observed regardless of a conservative approach, endovascular surgery, such as internal trapping or coil embolization assisted by stenting, becomes necessary.^{9,12,13}

In the present case, ischemic stroke occurred owing to dissecting aneurysm or BHS. The lesion was at the level of the

atlantoaxial joint, which was often stimulated repetitively by head rotation. We assumed that a conservative approach would be ineffective in preventing the recurrence of ischemic stroke and the worsening of dissecting aneurysm and VA, and thus we planned a surgical or endovascular approach.

There are many reports of BHS being caused by AAD,² but BHS caused by AAD with VA dissecting aneurysm is rare. To our knowledge, this is the first case report of BHS caused by AAD with VA dissecting aneurysm. After comparing endovascular and surgical approaches,

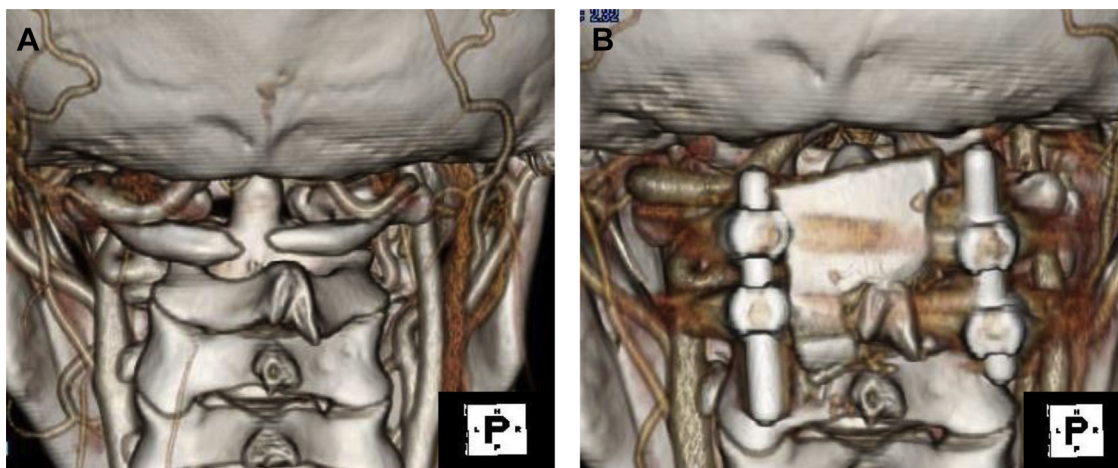


Figure 5. Three-dimensional computed tomography obtained preoperatively (A) and postoperatively (B). P indicates the posterior side of the patient.

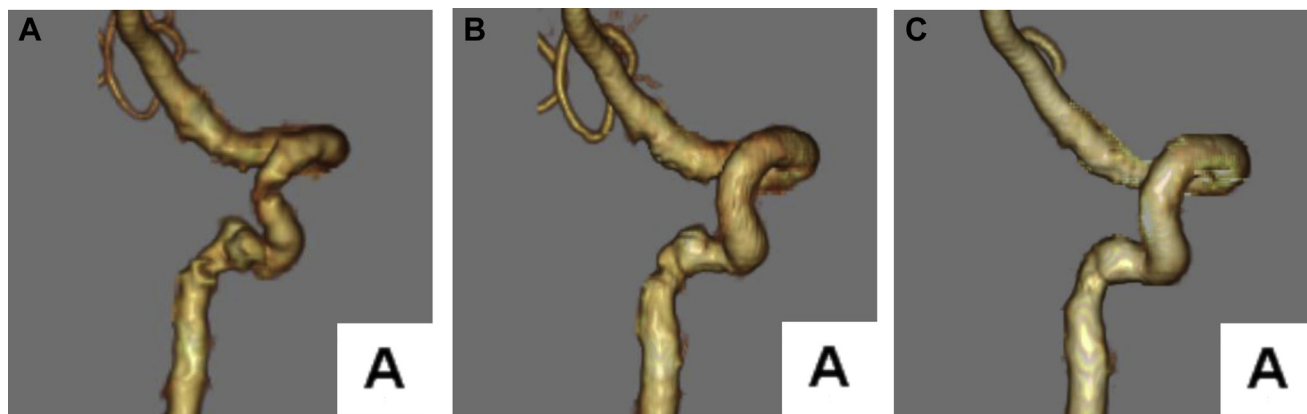


Figure 6. Three-dimensional computed tomography angiography (3D-CTA). (A) 3D-CTA obtained 7 days before surgery showed a left vertebral artery dissection at the V3 portion. (B) 3D-CTA obtained 5 months postoperatively showed the left vertebral artery dissection at the

V3 portion. (C) 3D-CTA obtained 8 months postoperatively showed shrinkage of the dissecting aneurysm. A indicates the anterior side of the patient.

we eliminated the endovascular approach for the following reasons. First, the endovascular approach using a stent could require antiplatelets for a long time. Thus, considering the patient's age, we avoided using antiplatelets. Second, we expected stent-assisted coil embolization would be needed for complete packing, and this involved a risk of sacrificing her dominant VA. Third, the endovascular approach was not curative for BHS and AAD. It was speculated that the dissecting aneurysm was caused by repetitive mechanical injury from the facet to the left VA. We postulated that C1-C2 posterior fusion was effective for BHS with VA dissecting aneurysm. Postoperatively, a second stroke did not occur. The patient had follow-up with computed tomography angiography for 8 months postoperatively; imaging showed that the dissecting aneurysm was growing smaller, and vessel wall irregularity and dilation were gradually improving. Thus, C1-C2 posterior fusion was effective for BHS with VA dissecting aneurysm.

CONCLUSIONS

We have presented a juvenile case of BHS caused by AAD with a VA dissecting aneurysm. Many cases of BHS caused by AAD have been reported, but to our knowledge this is the first case of BHS

with a VA dissecting aneurysm and aplasia at C1 lamina. Based on this case, we suggest that C1-C2 posterior fusion is effective for BHS with VA dissecting aneurysm.

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