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Case Report

Endovascular Therapy for an Aneurysm of a Distal Posterior Inferior Cerebellar Artery via the Ipsilateral Persistent Primitive Proatlantal Artery: A Case Report

Naoki Omura, Hiroto Kakita, and Fuminori Shimizu

Objective: We describe the rare case of a patient who was treated for a ruptured distal posterior inferior cerebellar artery (PICA) aneurysm via an ipsilateral persistent primitive proatlantal artery (PPPA).

Case Presentation: An 86-year-old female with a medical history of hypertension presented with headache and nausea. CT showed subarachnoid hemorrhage in the posterior cranial fossa, and CTA revealed an aneurysm at the left-side distal PICA. In the left-sided common carotid angiography, the artery with a branch of the occipital artery from the external carotid artery was described. This artery continued as the V3 segment and entered the dura via the foramen magnum. The artery perfused the territory of the left vertebral artery and PICA. We concluded that the artery, which entered the dura, was a PPPA. We decided to perform endovascular therapy that passed through the PPPA. The aneurysm was located in the cortical segments, beyond the cranial loop. We decided that parent artery occlusion (PAO) would be more effective than selective coil embolization to achieve safe and adequate hemostasis. The patient had a good outcome with PAO not assessing collateral circulation.

Conclusion: The emergency endovascular treatment with rare vascular variations requires accurate anatomical knowledge for treatment.

Keywords subarachnoid hemorrhage, distal posterior inferior cerebellar artery aneurysm, persistent primitive proatlantal artery, endovascular therapy

Introduction

Primitive carotid-vertebrobasilar anastomoses develop during the embryonic phase and aid in posterior cranial circulation. The four known types are the trigeminal, otic, hypoglossal, and proatlantal intersegmental arteries. These channels regressed in the week following the development of the posterior communicating artery. They rarely persist after birth or into adulthood, but are occasionally found incidentally due to intracranial vascular anomalies such as hemorrhages, aneurysms, and tumors. The persistent primitive proatlantal artery (PPPA) is one of these persistent carotid-vertebrobasilar anastomoses. It is rarely encountered clinically, with only one case found in 4400 cerebral angiograms.[1,2] A previous study has reported finding a PPPA unintentionally during endovascular treatment and utilizing it as an access route.[3]

Distal posterior inferior cerebellar artery (PICA) aneurysms are also unusual and are reported in 0.38%–1.7% cases.[2] Survivors of subarachnoid hemorrhage (SAH) due to distal PICA aneurysms generally have a good prognosis.[4]

Here, we report the case of an 86-year-old female who suffered from an SAH due to a ruptured aneurysm of the left distal PICA with an ipsilateral PPPA. The aneurysm was resolved with endovascular therapy.

Case Presentation

An 86-year-old female was admitted to the hospital, following a sudden onset of headache and nausea. The patient had a medical history of hypertension. On arrival, her

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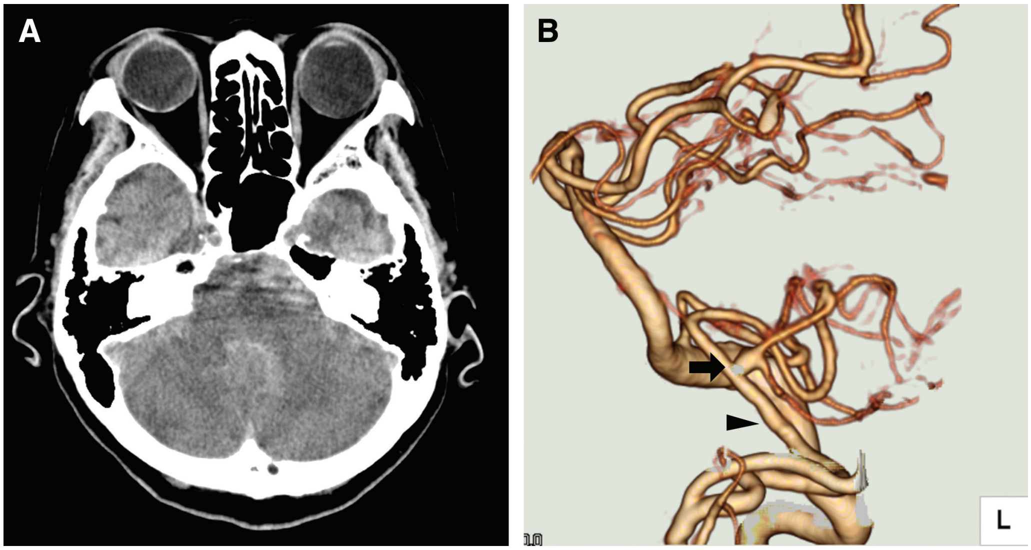

Fig. 1 Preoperative CT image and 3D-CTA of the patient. (**A**) Non-contrast CT depicting SAH in the posterior cranial fossa. (**B**) 3D-CTA from the left side. The PICA branched from the left VA (arrowhead). The ruptured aneurysm was found at the cortical segment of the left PICA (black arrow). PICA: posterior inferior cerebellar artery; SAH: subarachnoid hemorrhage; VA: vertebral artery

Glasgow Coma Scale score was 14 (E3V5M6). No neurological symptoms were observed. CT revealed SAH, primarily in the posterior cranial fossa (**Fig. 1A**). A three-dimensional CT angiogram revealed an aneurysm over the caudal loop of the left distal PICA (**Fig. 1B**). First, to diagnose and plan the endovascular treatment, we performed an angiography.

Angiography was performed under local anesthesia, via the right femoral sight, a 6-French (Fr) 25 cm long sheath (Medikit, Tokyo, Japan) and a 6-Fr 90 cm Fubuki guiding catheter (Asahi Intecc, Aichi, Japan). The origin of the left vertebral artery (VA) was not visible in the left subclavian artery angiogram (**Fig. 2A**). Imaging of the left common carotid artery (CCA) revealed an anastomosis between the ipsilateral carotid artery, the VA, and the aneurysm (**Fig. 2B**). This was an artery branching from the first segment of the common trunk of the occipital artery (OA), which continued as the V3 segment and entered the dura via the foramen magnum. The artery perfused the territory of the left VA and the PICA. A muscular branch of the OA was found at the horizontal segment. We judged the artery, which entered the dura, to be a PPPA. We decided to perform endovascular therapy that passed through the PPPA. In 3D-DSA, the aneurysm was located in the cortical segments, beyond the cranial loop. The maximum diameter and neck size of the aneurysm were 3.5 mm and 2.6 mm, respectively, and the size of PICA was 0.9 mm. Since the selective coil embolization was difficult and the aneurysm was located in the periphery, we decided that parent artery occlusion (PAO) would be more effective in achieving adequate hemostasis. Although the collateral circulation of this PICA had not been evaluated, PAO-induced neurological symptoms were expected to result in reversible ataxia, so the decision was made to perform PAO. PAO was performed by guiding an Excelsior SL-10 STR (Stryker, Kalamazoo, MI, USA) with a CHIKAI 10 microguidewire (Asahi Intecc) to the point just before the aneurysm (**Fig. 3B**). First, a Target 360 ULTRA 3 mm × 6 cm (Stryker) in dimensions was used as the first framing coil. The aneurysm was filled with 4 MicroPlex V-Trak10-HyperSoft 3D (Terumo, Tokyo, Japan) while kicking the microcatheter back. Finally, the proximal PICA of the aneurysm was occluded with 2-i-ED Coil Complex SilkySoft (Kaneka, Tokyo, Japan). In post-PAO imaging, the aneurysm and cortical segment of the PICA were not detected (**Fig. 3C**). The patient showed no neurological deficits after the treatment. Postoperative MRI showed only small ischemia in the left cerebellar hemisphere (**Fig. 3D**). The patient experienced no vasospasm or hydrocephalus and was discharged after 14 days of treatment. During a follow-up 3 months later, the patient visited the outpatient clinic and showed no neurological symptoms.

Discussion

The PPPA is a rare anastomosis between the carotid and vertebral arteries that is a remnant of embryonic circulation.[5] Yilmaz et al. reported only one case of persistent primitive

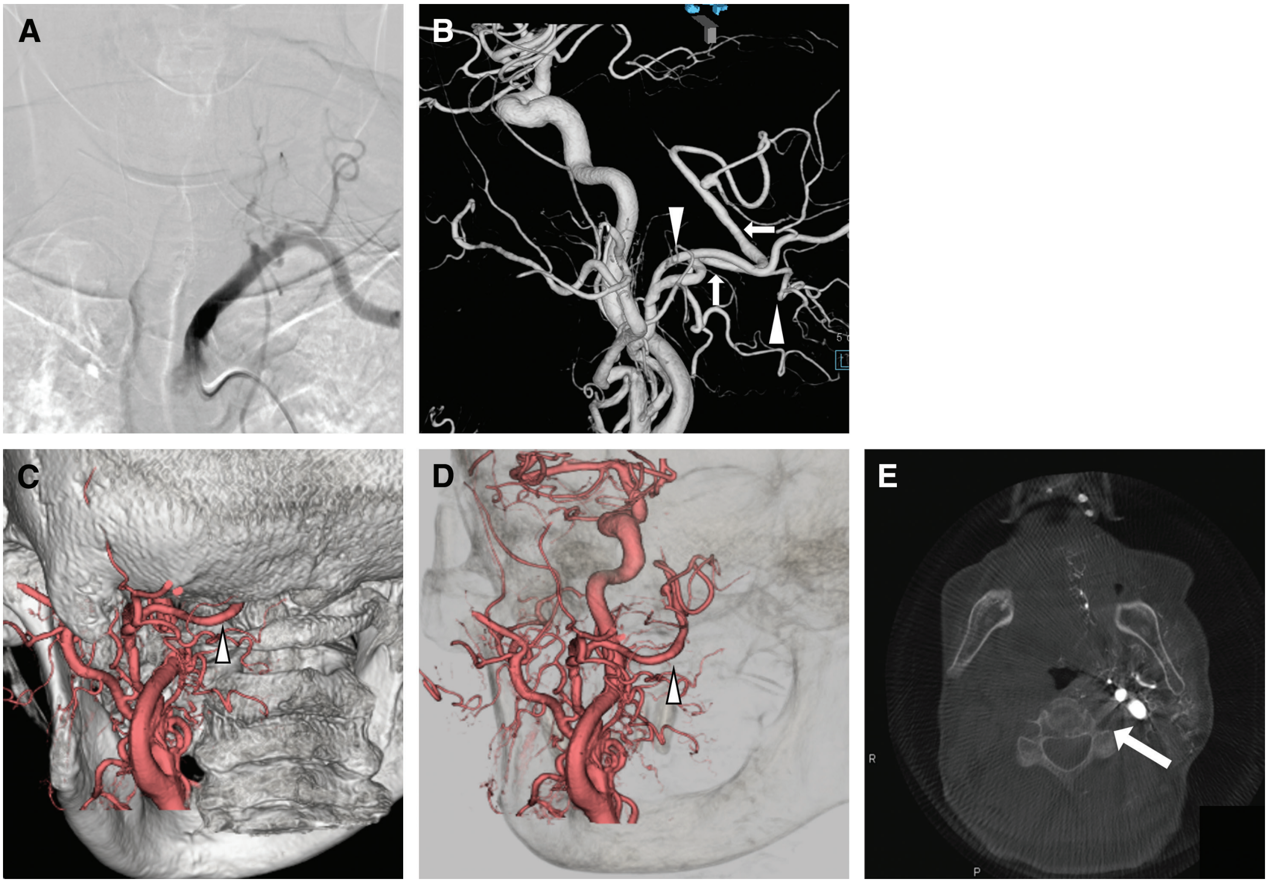

Fig. 2 (**A**) The origin of the left VA was not visible in the angiogram of the left subclavian artery. (**B**) 3D-DSA of the left CCA. A normal OA has muscular branches from its horizontal segment (arrowheads). The artery branching from the common trunk of the OA (arrow) connected with the VA. (**C** and **D**) 3D-DSA images of the patient with and without bones from the posterior view. The artery branched from the OA (arrowhead), ran over the atlas, and penetrated the foramen magnum. It did not pass through the C1 transverse foramen. (**E**) Axial slice of the third cervical vertebra in 3D-rotation angiography. The left-sided transverse vertebra was confirmed (arrow). No blood vessels were found passing through the foramen. CCA: common carotid artery; OA: occipital artery; VA: vertebral artery

carotid-vertebral anastomosis in 4400 cerebral angiograms.[1) The PPPA is classified into two types. Type I PPPA branches from the external carotid artery (ECA), internal carotid artery, or CCA. It does not pass through the transverse foramina of the cervical vertebrae. It connects with the V4 segment after passing the foramen magnum. Type II PPPA arises from the ECA and runs more laterally than type I. It passes through the transverse foramen of the C1 vertebra and joins with the V3 segment (**Fig. 4**). Although there are usually no branches arising from PPPA, OA can begin from PPPA.[6,7) The ipsilateral VA was found to be absent in 4 out of 21 patients with PPPA, as was also noted in this case.[8) Research indicates that 31.4% of PPPA-related aneurysms originate in the PPPA itself and 53% of them originate in the posterior of the circle of Willis.[9) There have been reports of endovascular treatment of aneurysms via PPPA and stent-assisted coiling for VA–PICA aneurysm by placing a stent through the contralateral PPPA.[3,10)

The OA has the potential for anastomosis with the VA via the meningeal collateral, the muscular branches, and direct anastomotic channels.[11) The frequency of visualized OA–VA anastomoses has been reported to be 14.9%.[12) Muscular branches of the OA may bifurcate downward from the horizontal part of the OA, draw loops, and form anastomoses with the VA. In our case, we identified diverging muscular branches of the OA and recognized that the artery arising from the common trunk of the OA was a PPPA. PPPA in the present case was similar to type I PPPA, but it did not pass through the C1 transverse foramen and was considered a subtype that branches OA.

Three-dimensional CT angiography of the transverse foramen of the C3 vertebra is useful in the differential diagnosis of PPPA to eliminate the possibility of chronic obstruction of the VA.[3) In PPPA, the transverse foramen is hypoplastic on the PPPA side. In the present case, the left-sided transverse foramen of the C3 vertebra was confirmed

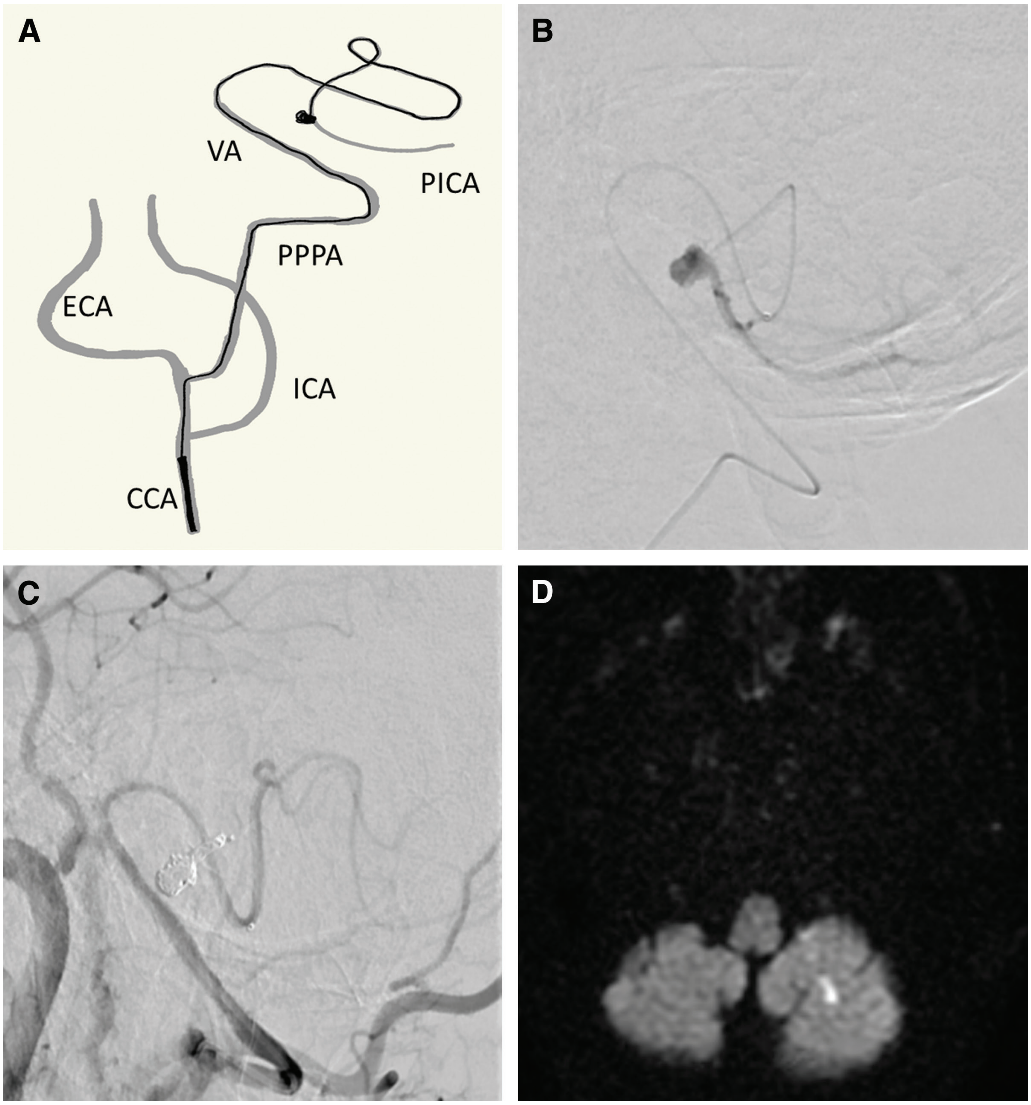

Fig. 3 (**A**) Schematic diagram of endovascular treatment. The guiding catheter was placed in the CCA. The microcatheter was guided to the aneurysm of the distal PICA via the PPPA. (**B**) Angiogram of the left PICA taken via a microcatheter before coil embolization. The tip of the microcatheter was guided to the point immediately before the aneurysm. (**C**) The aneurysm and the proximal artery were occluded with a coil. The aneurysm had disappeared in the post-embolization angiogram taken from the left CCA. (**D**) Postoperative MRI. Only spots with hyperintensity were apparent in the left cerebellar hemisphere. CCA: common carotid artery; PICA: posterior inferior cerebellar artery; PPPA: persistent primitive proatlantal artery

(**Fig. 2E**). However, no blood vessels were found passing through the foramen. In addition, the VA of cervical portion was not described retrogradely in the angiography of the left CCA.

Distal PICA aneurysms account for about 1.4% of all cerebral aneurysms and are often smaller and more fusiform than other aneurysms.[4)] The PICA is composed of five segments: the anterior medullary, the lateral medullary, the tonsillomedullary, the telovelotonsillar, and the cortical segments.[13)] The telovelotonsillar segment is the most frequent origin site of distal PICA aneurysms.[14)] There is no correlation between aneurysms and sides of the hypoplastic VA, and no aneurysm was generated on a hypoplastic PICA.[4)] Distal PICA aneurysms are frequently associated with other cerebrovascular diseases, such as brain arteriovenous malformation (AVM), which is most frequently reported. In the case of distal PICA aneurysm associated with small cerebellar AVM, CTA is insufficient, and DSA is required to evaluate nidus.[15)]

In the ruptured distal PICA aneurysm, the rebleeding rate is higher compared to that in other locations (43% vs. 21%); therefore, treatment for the ruptured distal PICA

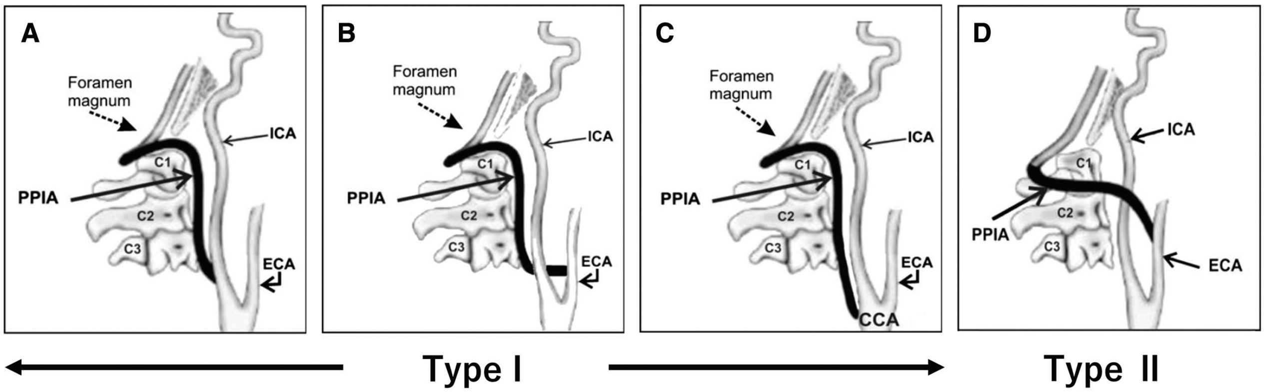

Fig. 4 Schematic of PPPAs (cited by reference 6). (**A**–**C**) The type I of PPPA branches from the ICA, ECA, or the CCA. It does not pass through the transverse foramina of the cervical vertebrae. It connects with the V4 segment after passing the foramen magnum. (**D**) Type II PPPA arises from the ECA and runs more laterally than type I. It passes through the transverse foramen of the C1 vertebra and joins with the V3 segment. CCA: common carotid artery; ECA: external carotid artery; ICA: internal carotid artery; PPPA: persistent primitive proatlantal artery

aneurysm should be given at the earliest.[4] The risk factors of unfavorable outcome are poor grade at admission and obstructive hydrocephalus in the patients with ruptured distal PICA.[2]

Microsurgical strategies for the treatment of distal PICA aneurysms are direct clipping,[16] wrapping, resection, or trapping with revascularization.[2] However, these treatments have the risk of brainstem and lower cranial nerve injury. The advantages of endovascular therapy for distal PICA aneurysms can help reduce the risk of posttreatment disorders in brain stem perforators and lower cranial nerves.

Chen et al. described the periphery of the distal PICA from the tonsillomedullary segment and reported that of the 36 patients with ruptured aneurysms of the distal PICA, 28 were treated with PAO and eight underwent selective aneurysm coiling (SAC).[17] PAO resulted in lower mortality (3.57% vs. 12.5%) but higher occlusion rates (100% vs. 87.5%) than SAC. Furthermore, 12 of the patients (33%) who underwent PAO experienced new cerebral infarctions, but none showed any new neurological symptoms. A study of 68 patients with SAH due to ruptured distal PICA aneurysms found that 52 patients recovered sufficiently to return to independent living, regardless of the surgical treatment modality (trapping, wrapping, proximal occlusion, or resection coagulation).[4]

In our case, we were able to perform PAO quickly, and the patient also returned to independent living.

Conclusion

Endovascular therapy for SAH resulting from a ruptured distal PICA aneurysm was successfully performed and resulted in a good clinical outcome. The emergency endovascular treatment with rare vascular variations requires accurate anatomical knowledge for safe treatment.

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Disclosure Statement

The authors declare that they have no conflicts of interest.

References

1) Yilmaz E, Ilgit E, Taner D. Primitive persistent carotid-basilar and carotid-vertebral anastomoses: a report of seven cases and a review of the literature. *Clin Anat* 1995; 8: 36–43.
2) Horiuchi T, Tanaka Y, Hongo K, et al. Characteristics of distal posteroinferior cerebellar artery aneurysms. *Neurosurgery* 2003; 53: 589–595; discussion 595-586.
3) Nariai Y, Takigawa T, Suzuki R, et al. Stent-assisted coil embolization for an unruptured vertebral artery-posterior inferior cerebellar artery aneurysm with retrograde access via type 1 persistent primitive proatlantal artery: a case report. *Interv Neuroradiol* 2021; 27: 397–401.
4) Lehto H, Harati A, Niemelä M, et al. Distal posterior inferior cerebellar artery aneurysms: clinical features and outcome of 80 patients. *World Neurosurg* 2014; 82: 702–713.
5) Hutchinson NA, Miller JD. Persistent proatlantal artery. *J Neurol Neurosurg Psychiatry* 1970; 33: 524–527.
6) Vasović L, Mojsilović M, Andelković Z, et al. Proatlantal intersegmental artery: a review of normal and pathological features. *Childs Nerv Syst* 2009; 25: 411–421.

7) Gumus T, Onal B, Ilgit ET. Bilateral persistence of type 1 proatlantal arteries: report of a case and review of the literature. *AJNR Am J Neuroradiol* 2004; 25: 1622–1624.
8) Choudhary G, Adhikari N, Chokr J, et al. Type 2 persistent primitive proatlantal intersegmental artery, a rare variant of persistent carotid-vertebrobasilar anastomoses. *Proc Bayl Univ Med Cent* 2019; 32: 101–104.
9) Kanematsu M, Satoh K, Nakajima N, et al. Ruptured aneurysm arising from a basilar artery fenestration and associated with a persistent primitive hypoglossal artery. Case report and review of the literature. *J Neurosurg* 2004; 101: 532–535.
10) Tian Y, Wang YF, Du HG, et al. Left type I proatlantal artery with bilateral aberrant vertebral arteries and a ruptured aneurysm: a case report and review of the literature. *Surg Radiol Anat* 2015; 37: 689–692.
11) Alvernia JE, Fraser K, Lanzino G. The occipital artery: a microanatomical study. *Neurosurgery* 2006; 58: ONS114–ONS122; discussion ONS114-ONS122.
12) Kajikawa R, Fujinaka T, Nakamura H, et al. Carotid artery stenting for patients with occipital-vertebral anastomosis. *Interv Neuroradiol* 2019; 25: 212–218.
13) Lister JR, Rhoton AL Jr., Matsushima T, et al. Microsurgical anatomy of the posterior inferior cerebellar artery. *Neurosurgery* 1982; 10: 170–199.
14) Lewis SB, Chang DJ, Peace DA, et al. Distal posterior inferior cerebellar artery aneurysms: clinical features and management. *J Neurosurg* 2002; 97: 756–766.
15) Case D, Kumpe D, Cava L, et al. Ruptured distal posterior inferior cerebellar artery (PICA) aneurysms associated with cerebellar arterial venous malformations (AVMS): a case series and review of the literature demonstrating the need for angiographic evaluation and feasibility of endovascular treatment. *World Neurosurg* 2017; 97: 751.e7–751.e13.
16) Orakcioglu B, Schuknecht B, Otani N, et al. Distal posterior inferior cerebellar artery aneurysms: clinical characteristics and surgical management. *Acta Neurochir (Wien)* 2005; 147: 1131–1139; discussion 1139.
17) Chen X, Sun Z, Shi L, et al. Endovascular management of ruptured distal posterior inferior cerebellar artery aneurysms: a retrospective cohort study. *Med (Baltim)* 2018; 97: e13300.