Dissection of the Posteroinferior Cerebellar Artery: Clinical Characteristics and Long-Term Follow-Up in Five Cases

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Key Words
Posterior inferior cerebellar artery · Dissecting aneurysm · Embolization · Subarachnoid hemorrhage

Abstract
Background and Purpose: Spontaneous isolated posteroinferior cerebellar artery (PICA) dissection is very rare. The aim of the study was to investigate the clinical manifestations, the neuroradiological aspects and the treatment of 5 cases collected in 5 years. Methods and Results: From 1999 to 2003, five patients (40–71 years old) were hospitalized for PICA dissection. Two patients presented symptoms after cervical manipulation. No predisposing factor or traumatic cause was described in the other cases. The diagnosis was carried out by cerebral angiography in all the cases: in 4, angiography showed focal stenosis with saccular or fusiform dilatation of the artery at the site of the dissection; in 1 a double lumen aspect was described. One patient presented ischemic manifestations; he was treated with heparin. The others presented subarachnoid hemorrhage (SAH) and were treated by embolization (endovascular sacrifice of the PICA). No ischemic complication and no rebleeding were observed after sacrifice of the PICA. Three patients presented acute hydrocephalus and were treated with surgical derivation. The angiographic and clinical follow-up lasted more than 3 years. We assessed the long-term results with the Glasgow Outcome Score and the modified Rankin Score. A good recovery was achieved for 4 patients (modified Rankin Score 0); one patient who presented SAH and who was in bad clinical state on admission had a Glasgow Outcome Score of 3 and a modified Rankin Score of 4 three years later. Conclusion: Clinical course and prognosis are variable in PICA dissections. The treatment depends on the existence of a hemorrhagic event. In the group of patients presenting SAH, endovascular treatment was safe and effective. The long-term clinical results depend mainly on the clinical status on admission.

Introduction
Spontaneous dissection of the posteroinferior cerebellar artery (PICA) is very rare, with only few cases reported in the literature [1–15]. Clinically the dissection may cause ischemic stroke or subarachnoid hemorrhage (SAH), depending on the dissecting plane of the affected artery. Five cases of PICA dissection were observed in 5 years in our institution; we present here clinical manifestations, angiographic aspects, prognosis, treatment and long-term follow-up in each of them.
Case Reports

Case 1
Mrs. P. is a 40-year-old woman who experienced a sudden headache during a sexual relation, and then she lost consciousness for a while. Hospitalized few hours later, she did not present any neurological deficit but a very intense headache and mild neck stiffness. Computed tomography (CT) scanning showed SAH predominating in the posterior fossa. Cerebral angiography revealed a 6-mm-large dissecting aneurysm of the right PICA. The origin of the PICA was extradural at the level of the first cervical vertebra (fig. 1), and the pseudoaneurysm, extracranial but intradural (fig. 1), was located below the foramen magnum on the ascending part of the PICA. Endovascular treatment was performed just after angiography: a microcatheter was advanced under fluoroscopic control in the right PICA; afterwards, 3 coils (Boston Scientific Corporation) were deployed in the pseudoaneurysm to occlude it. The patient’s postoperative course was unremarkable, and she was discharged without any neurological symptoms. Angiographic and clinical follow-up 1 and 3 years later showed a complete occlusion of the lesion and a complete recovery (fig. 2a, b).

Case 2
A 66-year-old man with a history of arterial hypertension developed a severe headache accompanied by nausea, vomiting and confusion. CT of the brain showed diffuse SAH with blood in the fourth ventricle and ventricular enlargement. Before cerebral angiography, an external ventricular drainage was performed. Cerebral angiography carried out at the time of the initial presentation was negative but performed in bad conditions because of a psychomotor agitation of the patient. An external ventricular drainage was placed and a second cerebral angiography was carried out 1 week later. This exam demonstrated a pseudoaneurysm on the tonsillar branch of the right PICA (fig. 3a, b). Endovascular treatment was performed at the same time; under general anesthesia, a 6-French guide catheter was placed in the right vertebral artery, and a microcatheter was advanced coaxially under fluoroscopic control in the right PICA just 1 cm before the pseudoaneurysm. Selective injection of contrast medium in the microcatheter showed an irregular aspect of the tonsillar artery and a 3-mm-large pseudoaneurysm (fig. 4). Embolization of the pseudoaneurysm and the dissected artery was performed with injection of a mixture of Histoacryl (Braun) and Lipiodol (Guerbet Medicale). The patient remained neurologically intact postoperatively and was discharged 3 weeks after. The neurological examination 3 years later was normal, and a cerebral angiography, carried out 3 months and 1 year after embolization, showed that the pseudoaneurysm was completely occluded with a patent right PICA (fig. 5).
Case 3
Mrs. T., a 46-year-old woman, presented vertigo and diplopia after a cervical manipulation. A few days later she experienced a severe headache with nuchal rigidity. A CT scan and a cerebral MRI were performed after hospitalization. These exams revealed an SAH in the posterior fossa (fig. 6a) and a cerebellar T2 hyper-signal corresponding to a cerebral ischemia in the territory of the right PICA (fig. 6b). A cerebral angiography showed an aspect of dissection of the lateromedullary segment of the right PICA (fig. 7a, b). Under general anesthesia, and via the transfemoral route, a microcatheter was advanced coaxially through a 6-French guiding catheter to the proximal part of the right PICA. Sacrifice of the dissected segment of the artery was performed with coils; the proximal part of the PICA, which may have perforators sup-
plying the brain stem, was preserved. The control angiogram obtained just after coiling showed opacification of the distal territory of the PICA thanks to the arterial anastomosis with the other cerebellar arteries. The neurological examination remained unchanged postoperatively. The patient was discharged 2 weeks later; she still presented a diplopia. Cerebral angiography 4 months later was similar to the control angiogram carried out after sacrifice of the PICA (fig. 8a, b). The clinical follow-up showed decreasing of the visual trouble in 1 year, and the patient presented no more symptoms after 2 years.

Case 4
Mrs. B., a 71-year-old woman, became unconscious while she was at home and fell on the floor. She was intubated and transferred to our institution for further care. Hypertension was the only medical problem known at this stage. Emergent cranial CT demonstrated subarachnoid and intraventricular hemorrhage and hydrocephalus. External ventricular drainage was performed. Cerebral angiography revealed a pseudoaneurysm on the trunk of the left PICA, 5 mm after its origin (fig. 9a, b). A microcatheter was advanced in the PICA. Occlusion of the pseudoaneurysm was performed with coils (fig. 10a, b). During her hospitalization, the clinical state improved slowly, and the patient was discharged to a nursing home 2 months after the hemorrhagic event. The clinical outcome was assessed every year. Three years after the hospitalization the patient presents intense weakness, diplopia and neurocognitive troubles. She is now classified as grade 3 in the Glasgow Outcome Score and grade 4 in the modified Rankin Classification.

Case 5
A few minutes after a session of cervical manipulation, Mrs. L., 42 years old, complained of intense posterior headache, dizziness and vomiting. The patient was admitted to hospital the same day. Initial clinical examination showed painful movement of the cervical spine, rotating dizziness and a cerebellar syndrome associating both ataxia and dysmetria. There was no motor deficit or impairment of sensation and no impairment of the last cranial nerves. A CT scan of the brain, carried out the day after her admission, revealed a hypodensity of the right cerebellar region, suggesting a stroke.

A Doppler exam demonstrated good permeability of the vertebral arteries. Cerebral angiography was carried out with catheterization of both vertebral arteries. Both vertebral arteries ap-
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Appeared normal. The right PICA showed an extracranial origin. One centimeter after its origin, we observed a 5-mm-long arterial segment with an irregular appearance of the wall and a double lumen aspect suggesting arterial dissection (fig. 11). Moreover, angiography revealed a considerable slowdown of flow in this artery. Intravenous administration of heparin was implemented for 2 weeks, during which the patient’s clinical state improved considerably, with disappearance of dizziness and vomiting. The patient left the hospital on the 20th day; she was able to walk, but residual mild headaches as well as neck stiffness were experienced. After 2 years, the patient was free of symptoms.

Discussion

Dissections of intracranial arteries in the posterior circulation most commonly present with SAH and more frequently occur on the vertebral artery. Isolated dissection of the PICA is rare [1–15]. Although clinical manifestations may be ischemic or hemorrhagic, most of the
reported cases in the literature presented SAH with dissecting aneurysms. Contrary to dissections with hemorrhagic manifestations, the diagnosis of dissection with ischemic symptoms is probably underestimated because cerebral angiography is generally not performed in this case. This predominance of the hemorrhagic type was also observed in our series: 3 patients presented SAH, 1 presented ischemic symptoms and 1 both hemorrhagic and ischemic symptoms.

Although 2 groups of patients seem to be distinguished, some overlap may occur like in our third case. Progression from the ischemic type to the hemorrhagic type is possible for Gaillard et al. [16]. For these authors, subsequent angiographic imaging must be performed in the ischemic type to search for pseudoaneurysms which may require specific treatment.

The age of onset is variable, from 20 to 71 years, with predominance in the fourth decade of life [2]. Men and women are equally affected [2, 5, 11]. Every segment of the PICA can be affected, but the first segment of the artery, the anterior medullary segment, is the most frequently affected one. For Yamakawa et al. [11], dissection located in the first segment of the PICA would tend to cause ischemic events, whereas more distally located ones would tend to give rise to hemorrhagic events. On the contrary, Kanou et al. [3] noted that dissecting aneurysms of the proximal PICA tend to cause infarctions and that peripheral ones tend to lead to SAH.

Although to most of the authors the cause of PICA dissection is unknown, 3 cases of dissection associated with segmental arteriopathy have been described in the literature [11, 14]. Some authors have suggested that hypertension may play an underlying role [12, 13]. One case of PICA dissection has been reported after direct trauma (transoral tumor biopsy) [15]; in our series, the onset of the symptoms began after cervical manipulation in 2 cases.

CT and MR imaging may show subarachnoid/intraventricular hemorrhage and/or infarction, but cerebral angiography is the ‘gold standard’ imaging modality for the diagnosis of dissection; pearl and string aspect is the most frequent sign with generally a smooth segmental tapered stenosis followed by a fusiform or saccular dilatation of the artery. Segmental narrowing and occlusion of the vessel have been described; the pathognomonic ‘double lumen’ aspect, observed in one of our cases (fig. 11), is very rare. Because of the small caliber of the PICA, fusiform dilatation with or without a saccular pouch corresponding to a pseudoaneurysm seems to be the most specific aspect; a clinicopathological study of dissecting aneurysms of the intracranial arteries has shown that this angiographic aspect is dominant in SAH cases, whereas stenosis without dilatation is dominant in ischemic cases [17]. Arterial dissections are not always obvious on initial angiograms. An acute awareness of intracranial dissection as a source of bleeding is warranted in evaluating patients with ‘angiogram-negative’ SAH; a second angiogram carried out few days after is sometimes justified; above all, if the first angiography has not been performed in good condition, like in our second patient (fig. 3a, b).

In the ischemic cases, the prognosis of PICA infarction is generally good [18, 19] except for patients with multiple brainstem lesions [20]; in the SAH cases, the prognosis of vertebrobasilar dissections is usually poor with regard to the initial state, and there is a high risk of rebleeding [21, 22]. Dissecting aneurysm of the PICA carries a 24% risk of rebleeding in the acute phase, associated with a high mortality rate [23]. Presence of a pseudoaneurysm substantially increases the risk of early rebleeding [24, 25] and early intervention is essential. Even though early intervention seems essential to avoid rebleeding, the prognosis of these lesions is mainly correlated with the clinical state on admission. High clinical grades with trouble of consciousness are strongly predictive of poor outcome [25].

**Treatment**

In the ischemic cases, little is known about the risk of hemorrhagic rupture and there is no consensus on management. Although some authors [26] have written that patients with intracranial dissections with ischemic symptoms are also at risk for subsequent SAH, conservative management with medical therapy is considered as the most appropriate therapeutic plan, above all, if no pseudoaneurysm has been observed on the angiogram. This medical treatment includes anticoagulants or antiplatelet drugs, but no randomized trials or reliable comparisons of the two treatments are available. In these unruptured dissections, early angiographic control must be performed to search for pseudoaneurysm [16] because such a condition may be amenable to surgical or endovascular treatment.

In the SAH cases, the use of a conservative approach with anticoagulation, such as the one used in arterial dissection without SAH, is problematic. In the literature the conservative treatment has been reserved for patients in very poor clinical conditions. For the other patients, the goal of the (surgical or endovascular) treatment is to occlude the dissected arterial segment and especially the pseudoaneurysm to prevent rebleeding during the acute phase.
The optimal treatment of intracranial dissecting aneurysm is subject to controversy [5, 7, 10, 23]. Most of the patients in the literature have been treated surgically [1, 2, 6, 8, 9, 11, 27]. Surgery includes proximal clipping, trapping and wrapping or resection with PICA end-to-end anastomosis, PICA-to-occipital artery anastomosis or PICA reimplantation to the vertebral artery. However, manipulating the proximal PICA for clipping has a high risk of postoperative neurological morbidity because of the close relationship between the brain stem and lower cranial nerves [27, 28].

The endovascular treatment consists in occluding the dissected site with coil or glue. This treatment [7, 10, 12, 24] has shown effectiveness to avoid rebleeding [7, 10, 12], and because endovascular procedures do not involve manipulation of the brain stem, cranial nerves or cerebellar tissue, the complications noted previously may be avoided.

Surgical or endovascular trapping of the dissected arterial segment may lead to 2 main complications: an infarct in the distal territory of the PICA and an ischemic lesion of the medulla due to occlusion of perforators in the trapped segment.

Apart from the caudal medulla, the PICA supplies blood to the cerebellar tonsils, inferior portion of the cerebellar hemisphere, vermix and choroid plexus of the fourth ventricle: sacrifice of the PICA can potentially result in neurological deficit, and surgical anastomosis could be considered essential. However, anatomical studies and surgical experience indicate that for most patients with PICA dissection, good collateral flow via the anterior inferior cerebellar artery (AICA) and the superior cerebellar artery exits, and that vascular anastomosis to supply the area distal to the trapped segment is unnecessary [2]. Moreover, the occipital artery to PICA bypass before trapping is not feasible in all cases due to the small size of the PICA or other technical difficulties. Finally, the effect of the superficial cortical infarct produced by occlusion of the distal PICA is usually not clinically significant [2]. In fact, it seems essential to stress the numerous anatomical variations in the vascular anatomy of the PICA. This can explain why it is difficult to recommend a standardized approach. For Maimon et al. [12], cerebral angiography can distinguish between 2 types of anatomy. In the first one the ipsilateral (AICA) is greater than, equal to or more than half of the size of the PICA: in this case endovascular occlusion of the vessel would not be associated with significant sequelae. The second disposition includes a small AICA and a dominant PICA or AICA/PICA variant: in this type, Maimon et al. [12] suggest performing a test occlusion with careful consideration of alternative surgical treatment.

A more frightening complication in the endovascular treatment is the sacrifice of the perforating vessels. Lister et al. [29] studied PICA anatomy in 25 adult cadavers. They reported that the perforating branches emanated from the first 3 segments of the PICA (anterior medulary, lateral medullary, tonsillomedullary) and especially from the first one. Thus, if the pseudoaneurysm is located distally, a sacrifice of the PICA distal to its third segment (approximately halfway between the caudal and rostral loops of the PICA [2]) will be usually well tolerated. For Lasjaunias and Berenstein [30], the perforators from the PICA do not depend on the size of the artery but on where the origin of the cerebellar artery is positioned; the higher the origin of the PICA, the higher the chances of it having medullary branches. Therefore, PICA with an extracranial origin can be also sacrificed safely.

For Yamakawa et al. [11], intraoperative inspection of the perforating branches helps assess the need for revascularization. On the contrary, Maimon et al. [12] stated that distal anastomoses from the other cerebellar arteries do nothing to compensate for the loss of perforators in the segment of the PICA that is trapped. It seems important when the dissection is located in the first 3 segments of the PICA, and especially in the first one, to carefully select the arterial segment to occlude to minimize damage to the perforators.

References


