Isolated Basilar Artery Dissection with Ischemic Stroke: Report of 4 Cases

Szu-Ju Chen[1,2], Li-Kai Tsai[1], Sung-Chun Tang[1], Jiann-Shing Jeng[1]

Abstract

Purpose: Isolated basilar artery dissection (BAD) is a rare cause of ischemic stroke. Since the clinical presentations and imaging findings could be non-specific and subtle, the diagnosis may be difficult. Here, we presented four cases of isolated BAD with acute ischemic stroke.

Case report: Four patients (age, 22 to 57 years) experienced acute onset of hemiparesis and/or vertigo with half of them having initial headache. Acute infarct was noted at pons or thalamus. Three cases needed more than one imaging modality or serial follow-up imaging to confirm a diagnosis of isolated BAD. Vascular tapering and/or false lumen restrictive to basilar artery were the commonest imaging findings. Three of our patients received anticoagulant without recurrent infarct or hemorrhagic complication. All of the patients had good functional outcome with modified Ranking scale scoring 1.

Conclusion: Isolated BAD may cause variable clinical manifestations and the outcome can be favorable. Application of different and advanced imaging studies with serial image follow-up are useful to confirm the diagnosis.

Key Words: arterial dissection, basilar artery, ischemic stroke, neuroimaging

Acta Neurol Taiwan 2017;26:138-143

INTRODUCTION

Isolated basilar artery dissection (BAD) is a rare but important cause of ischemic stroke. The annual incidence was estimated at 1/400,000[(1)]. Clinical manifestations of isolated BAD include ischemic stroke or transient ischemic attack, subarachnoid hemorrhage as a consequence of dissecting aneurysm rupture, and brainstem compression by large pseudoaneurysm, etc[(1)]. Since neurological events often occurred within 1 week after artery dissection and the risk of recurrent stroke is highest in the first month[(2,3)], early diagnosis and treatment are important to avoid mortality and morbidity.

Currently, there is no consensus diagnostic criteria or treatment guideline for isolated BAD. The difficulty in diagnosing isolated BAD arises from its widely variable clinical presentations, and subtle and non-specific radiological signs on imaging studies[(3)]. Hereby, we presented four cases of isolated BAD to share our diagnostic processes and treatment results (Table 1).

From the Departments of [1]Neurology, National Taiwan University Hospital, Taipei, Taiwan; and [2]Neurology, En Chu Kong Hospital, New Taipei City, Taiwan.
Received July 24, 2017.
Revised and Accepted November 27, 2017.
Correspondence to: Li-Kai Tsai, MD, PhD. Department of Neurology, National Taiwan University Hospital, No. 7, Chung-Shan South Road, Taipei 100, Taiwan.
E-mail: 009502@ntuh.gov.tw

Table 1. Clinical manifestations, treatments and outcomes of our four patients

Age/sex	Clinical presentation	Radiological findings	Treatment	Outcome
57M	slurred speech, left hemiparesis	MRI: pontine infarct and severe stenosis/ occlusion at BA with tapering lumens, intimal flap, double lumen and intramural hematoma.	dual antiplatelet therapy (aspirin plus clopidogrel) for 15 months	NIHSS 0 at discharge; mRS 1 at post-stroke 3 months
36M	vertigo, dizziness, vomiting, right hand numbness and clumsiness	MRI: multiple infarctions in left pons with BA occlusion. CTA: BA occlusion with pearl and string sign.	anticoagulant therapy for 6 months	NIHSS 0 at discharge; mRS 1 at post-stroke 3 months
39F	headache, vertigo, transient loss of consciousness, right hand clumsiness, right side numbness	MRI: subacute bilateral thalamic hemorrhage and severe BA stenosis with tapering lumens. CTA at post-stroke 3 months: complete recanalization of BA.	anticoagulant therapy for 4 months	mRS 1 at post-stroke 3 months
22F	occipital headache, left hemiparesis, vertigo, nausea, vomiting, transient blurred vision	CTA and conventional angiography: focal filling defects at BA. High resolution MRI: an intimal flap with double lumen and an intramural hematoma at BA.	anticoagulant therapy for 5 months	NIHSS 0 at discharge; mRS 1 at post-stroke 3 months

CASE PRESENTATION

Case 1

A 57-year-old man suffered from acute onset of slurred speech and left hemiparesis. No headache or vertigo was noted. Brain MRI on the second day showed acute right paramedian pontine infarct on diffusion weighted imaging (DWI). MRA study showed occlusion or severe stenosis at the basilar artery (BA) with tapering lumen (Fig. 1A). T2-weighted imaging (T2WI) demonstrated an

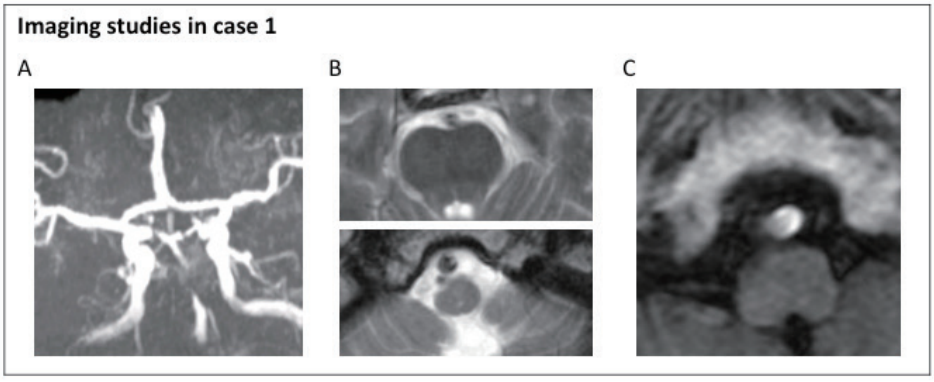

Figure 1. The neuroimaging of case 1. (A) MRA showed severe basilar artery (BA) stenosis with tapering lumens. (B) T2WI demonstrated an intimal flap and double lumen in BA. (C) T1WI revealed an intramural hematoma at BA presenting with crescentic hyperintense lesion.

intimal flap with double lumens within the BA (Fig. 1B). T1-weighted imaging (T1WI) visualized a hyperintense crescent lesion in the BA, indicating intramural hematoma (Fig. 1C). Acute ischemic stroke caused by isolated BAD was diagnosed and he was treated with dual antiplatelet therapy, aspirin plus clopidogrel, for 15 months. Follow-up CTA study 3 months after stroke still showed severe BA stenosis. The patient did not experience neurological deterioration with NIHSS score of 0 at discharge and modified Rankin Scale (mRS) score of 1 at 3 months post-stroke.

Case 2

A 36-year-old man suffered from transient vertigo, followed by acute onset of the right hand numbness, dizziness, and vomiting 3 hours later. Initial non-contrast brain CT showed unremarkable findings. However, clumsy right hand developed further. Neurological examination showed hypoesthesia and ataxia in the right limbs with a NIHSS score of 2. Brain MRI study revealed multiple infarcts in the left pons with mixed acute and subacute stages. MRA depicted BA occlusion (Fig. 2A) and CTA showed BA occlusion with pearl and string sign (Fig. 2B), leading to a diagnosis of isolated BAD. Oral anticoagulant therapy was used for 6 months. Follow-up CTA at 3-month post-stroke still showed BA occlusion. His neurological condition remained stable with NIHSS score of 0 at discharge and mRS score of 1 at 3 months post-stroke.

Case 3

A 39-year-old woman suffered from acute onset of headache and vertigo, followed by transient loss of consciousness for 20 minutes. Four days later, she developed acute onset of clumsy right hand and right hemi-tingling sensation. Serial work-up did not reveal any abnormality until brain MRI performed 3 weeks later, which showed subacute intracranial hemorrhage in the right medial and left posterior thalamus. MRA revealed severe BA stenosis with tapering lumen (Fig. 3A). Isolated BAD with bilateral thalamic embolic infarct, followed by hemorrhagic transformation was suspected. She received anticoagulant smoothly for 4 months without neurological deterioration. Follow-up CTA at post-stroke 3 months showed complete recanalization of BA (Fig. 3B), which further confirmed a diagnosis of BAD. Her mRS score was 1 at 3 months post-stroke.

Case 4

A 22-year-old woman developed acute onset of left hemiparesis preceded by occipital headache with nausea, vomiting, transient blurred vision and vertigo. Initial NIHSS score was 3 and improved to 1 within one day. Non-contrast head CT, brain MRI/MRA, and cervical MRI did not reveal any significant abnormality. CTA and conventional angiography showed focal filling defects at the BA (Fig. 4A). High resolution brain MRI with contrast enhancement showed an intimal flap with

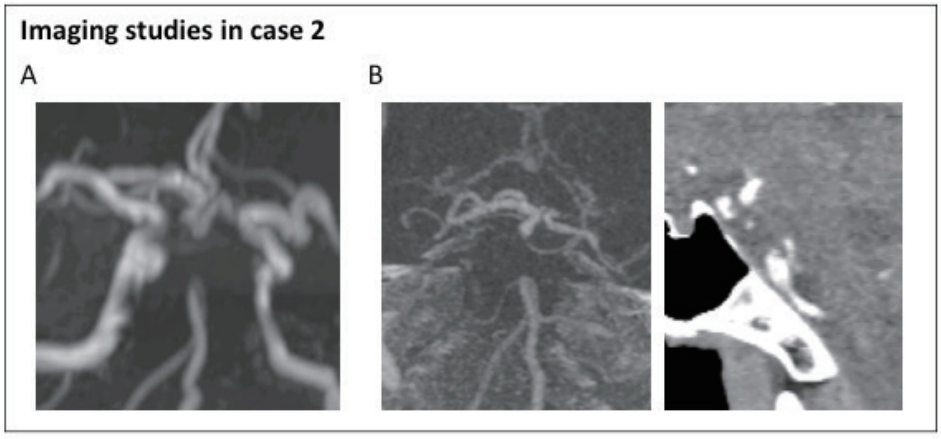

Figure 2. The neuroimaging of case 2. (A) MRA depicted BA occlusion. (B) CTA showed BA occlusion with pearl and string sign.

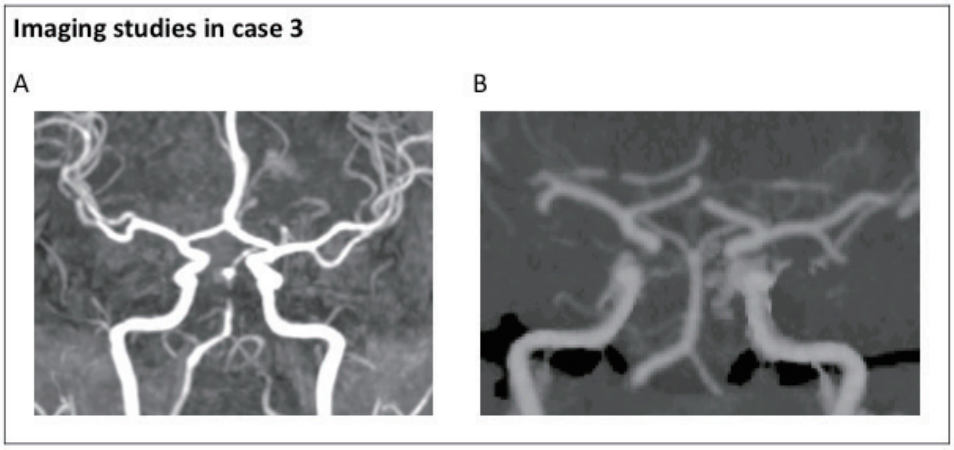

Figure 3. The neuroimaging of case 3. (A) MRA revealed severe BA stenosis with tapering lumen. (B) CTA performed 3 months later showed re-canalization of BA.

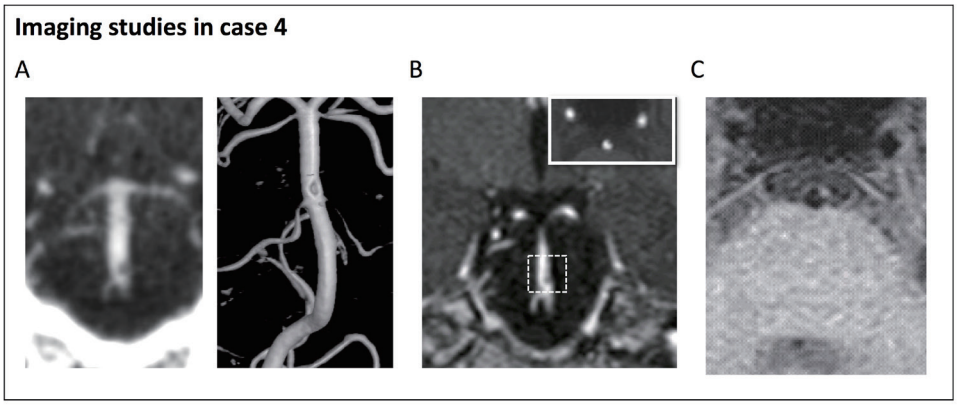

Figure 4. The neuroimaging of case 4. (A) CTA and conventional angiography revealed focal filling defects at BA, indicating focal stenosis. (B) High resolution post-gadolinium contrast enhanced T1WI showed an intimal flap at BA. (C) High resolution axial T1WI visualized an intimal flap with double lumen sign and an intramural hematoma presenting with hyperintense lesion in the vessel wall at the left portion of the flap.

hyperintense intramural hematoma at BA by T1WI (Fig. 4 B,C), which supported a diagnosis of isolated BAD. The patient received oral anticoagulant for 5 months without neurological worsening. Follow-up high resolution MRI 3 months later still showed a thin flap in the BA. Her NIHSS score was 0 at discharge and mRS was 1 at 3 months post-stroke.

DISCUSSION

Isolated BAD is an increasingly identified cause of cerebrovascular disease. Clinical manifestation of BAD differs according to the territory of infarction or hemorrhage and the degree of compression by dissected vessel walls. Hemiparesis, dysarthria, nausea/vomiting, vertigo, and sudden consciousness change were frequently recorded symptoms in isolated BAD[1,4,5]. By contrast, Wallenberg syndrome was reported to be the most common presented clinical syndrome in intracranial vertebral artery (VA) dissection[6]. Headache and neck pain are cardinal features of artery dissection and were reported in 18.9 to 92% of patients in intracranial artery dissection[1,3,5-7]. In our cases, hemiparesis and vertigo were the most common initial symptoms and one patient had a

transient consciousness change. Headache only occurred in two of four patients; therefore, artery dissection should not only be suspected in patients with headache or neck pain.

In our cases, the most common mechanism of ischemic stroke caused by isolated BAD was occlusion of the local BA branches. The result was supported by one previous study showing that local branches occlusion was the main stroke mechanism for isolated BAD (75%) and intracranial VA dissection (75%)[8]. In contrast, the most frequent mechanism of cerebral infarct is artery-to-artery embolism in patients with extracranial arterial dissection or intracranial artery dissection of the anterior circulation[8]. It may be because the perforating vessels of BA and VA are shorter and smaller than those in anterior circulation, which makes them more vulnerable to occlusion[8].

Although conventional angiography remains the gold standard for diagnosing artery dissection, the invasiveness impedes its regular use[2,9,10]. In clinical practice, MRI, MRA and CTA are the first-line imaging studies in patients suspected of isolated BAD. However, the pathognomonic radiological findings of BAD are relatively uncommon, such as intramural hematoma, intimal flap or double lumens[2,9-12]. Other classical findings are often required to confirm the diagnosis of BAD, including arterial narrowing with distal dilation (pearl sign), narrowing of the artery lumen (string sign), tapered occlusion, or recanalization of previous occluded/stenosed arteries noted during follow-up[2,9-11]. In recent years, the application of high-resolution MRI has provided better sensitivity of visualizing intramural hematoma, short-segment dissection and lesions in tortuous or small-diameter vessels thereby increased the diagnostic rate of isolated BAD[1,2,10]. Only one of our four cases got a diagnosis of isolated BAD using the first-line imaging study. The other three cases needed to receive more than one imaging modality or serial imaging studies to confirm the diagnosis of BAD. In case 4, only high-resolution MRI was able to depict an intimal flap and intramural hematoma. Since the arterial lesions in isolated BAD could be short and hard to be detected, using different imaging modalities, more advanced techniques, and serial follow-up imaging could provide more radiological evidences for diagnosis of isolated BAD.

Currently, there are still no randomized trials to guide the optimal treatment for isolated BAD. In clinical practice, endovascular or surgical treatments are usually reserved for patients with ruptured aneurysm, compressive neurological deficits, or recurrent stroke despite adequate medical treatment[1,9,13]. While usage of anti-thrombotic agents is usually considered as regular treatment for patients with cervical or intracranial arterial dissection, whether anticoagulants or antiplatelet agents have better efficacy in prevention of recurrent stroke is still under debate[1,8,9,13]. For isolated BAD, three of our four cases received anticoagulants in acute and/or subacute stages for four to six months without further recurrent embolic or hemorrhagic insults and all their neurological outcome is favorable. A previous study also reported that 5 out of 7 BAD patients receiving anticoagulant treatment showed good clinical outcome without hemorrhagic events[1]. Notably, BAD was known to have poor prognosis with a mortality rate ranging from 10% to 78.9%[13]. In patients with BAD, thrombotic propagation in false lumen may diminish blood flow in penetrating branches of BA and also increase the risk of thromboembolism[1,8,9]. In addition, cerebral infarct in the posterior fossa is less vulnerable to develop hemorrhagic transformation[14]. Therefore, anticoagulants might be a reasonable choice for patients with BAD complicated with ischemic stroke but without aneurysm development.

In conclusion, isolated BAD may cause variable clinical manifestations and the outcome can be favorable. Application of different and advanced imaging studies with serial image follow-up are useful to confirm the diagnosis. Although the optimal treatment for BAD is still unclear, the use of anticoagulants may be safe and effective to prevent further embolic infarct.

REFERENCE

1. Ruecker M, Furtner M, Knoflach M, Werner P, Gotwald T, Chemelli A, Zangerle A, Prantl B, Matosević B, Schmidauer C, Schmutzhard E, Willeit J, Kiechl S. Basilar artery dissection: Series of 12 consecutive cases and review of the literature. Cerebrovas Dis 2010;30:267-276.
2. Han M, Rim N, Lee JS, Kim SY, Choi JW. Feasibility of high-resolution MR imaging for the diagnosis of intracranial vertebrobasilar artery dissection. Eur

Radiol 2014;24:3017-3024.
3. Debette S, Compter A, Labeyrie MA, Uyttenboogaart M, Metso TM, Majersik JJ, Goeggel-Simonetti B, Engelter ST, Pezzini A, Bijlenga P, Southerland AM, Naggara O, Béjot Y, Cole JW, Ducros A, Giacalone G, Schilling S, Reiner P, Sarikaya H, Welleweerd JC, Kappelle LJ, de Borst GJ, Bonati LH, Jung S, Thijs V, Martin JJ, Brandt T, Grond-Ginsbach C, Kloss M, Mizutani T, Minematsu K, Meschia JF, Pereira VM, Bersano A, Touzé E, Lyrer PA, Leys D, Chabriat H, Markus HS, Worrall BB, Chabrier S, Baumgartner R, Stapf C, Tatlisumak T, Arnold M, Bousser MG. Epidemiology, pathophysiology, diagnosis, and management of intracranial artery dissection. Lancet Neurol 2015;14;640-654.
4. Caplan LR. Dissections of brain-supplying arteries. Nat Clin Prac Neurol 2008;4:34-42.
5. Tsao Y. Isolated basilar artery dissection--a rare cause of stroke in young adult. Am J Emerg Med 2013; 31:1422.e3-5.
6. Hosoya T, Adachi M, Yamaguchi K, Haku T, Kayama T, Kato T. Clinical and neuroradiological features of intracranial vertebrobasilar artery dissection. Stroke 1999;30:1083-1090.
7. Chang FC, Yong CS, Huang HC, Tsai JY, Sheng WY, Hu HH, Chung CP. Posterior circulation ischemic stroke caused by arterial dissection: characteristics and predictors of poor outcomes. Cerebrovasc Dis 2015;40:144-150.
8. Kwon JY, Kim N-Y, Suh DC, Kang D-W, Kwon SU, Kim JS. Intracranial and extracranial arterial dissection presenting with ischemic stroke: lesion location and stroke mechanism. J Neurol Sci. 2015;358:371-376.
9. M.S. Ali, P.S. Amenta, R.M. Starke, P.M. Jabbour, L.F. Gonzalez, S.I. Tjoumakaris, A.E. Flanders, R.H. Rosenwasser, A.S. Dumont. Intracranial Vertebral Artery Dissections: Evolving Perspectives. Intervent Neuroradiol 2012;18:469-483.
10. Obusez EC, Jones SE, Hui F. Vessel wall MRI for suspected isolated basilar artery dissection. J Clin Neurosc 2016;27:177-179.
11. Gottesman RF, Sharma P, Robinson KA, Arnan M, Tsui M, Saber-Tehrani A, Newman-Toker DE. Imaging characteristics of symptomatic vertebral artery dissection: a systematic review. Neurologist 2012;18:255-260.
12. Ahn SS, Kim BM, Suh SH, Kim DJ, Kim DI, Shin YS, Ha SY, Kwon YS. Spontaneous symptomatic intracranial vertebrobasilar dissection: initial and follow-up imaging findings. Radiology 2012;264:196-202.
13. Li L, Li T, Xue J, Wang Z, Bai W, Zhu L, Feng G, Xu G, Yang B. Stent treatment for basilar artery dissection: A single-center experience of 21 patients. Interv Neuroradiol 2016;22:260-265.
14. Dorňák T, Král M, Hazlinger M, Herzig R, Veverka T, Buřval S, Šaňák D, Zapletalová J, Antalíková K, Kaňovský P. Posterior vs. anterior circulation infarction: demography, outcomes, and frequency of hemorrhage after thrombolysis. Int J Stroke 2015;10:1224-1228.