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Diagnosis of Intracranial Artery Dissection

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Abstract

Cerebral arterial dissection is defined as a hematoma in the wall of a cervical or an intracranial artery. Cerebral arterial dissection causes arterial stenosis, occlusion, and aneurysm, resulting in acute infarction and hemorrhage. Image analysis by such methods as conventional angiography, computed tomography, magnetic resonance imaging, and so on plays an important role in diagnosing cerebral arterial dissection. In this study, we explore the methods and findings involved in the diagnosis of cerebral arterial dissection.

Key words: cerebral arterial dissection, CT, MRI

Introduction

Cerebral arterial dissection is defined as a hematoma in the wall of a cervical or an intracranial artery. Cerebral arterial dissection causes arterial stenosis, occlusion, and aneurysm, leading in turn to brain infarction and subarachnoid hemorrhage. Arterial dissection may occur in any artery,[1,2] but it occurs predominantly in the posterior circulation in Japan, and predominantly in the anterior circulation in western countries.[3] Of particular importance is its relation to the lateral medullary infarction known as Wallenberg syndrome.[4–10] Imaging is acknowledged to play a central role in the diagnosis of cerebral arterial dissection; we would even say that it is the only acceptable method of diagnosis. The imaging techniques used for this purpose are conventional angiography, computed tomography (CT), and magnetic resonance imaging (MRI). Diagnosis requires the presence of intramural hematoma and dissecting aneurysm as demonstrated through image analysis. In this study, we have explored the methods and findings involved in the diagnosis of cerebral arterial dissection.

Features of Cerebral Arterial Dissection

Cerebral artery dissection is encountered in many hospitals. At our hospital, we have treated 107 patients with cerebral arterial dissection (including strongly suspicious and clinically suspicious cases) between January 1997 and July 2015 (71 men and 36 women; mean age, 59.2 ± 14.8 years). Cerebral arterial dissection occurs frequently in younger patients. In our series, 24 patients (22%) were in the second, third or fourth decades of life. Onset was acute in 68% of our cases. Clinical symptoms were noted in 63 cases with infarction (including 14 cases with Wallenberg syndrome) and 13 patients with intracranial hemorrhage (12 patients with subarachnoid hemorrhage and one patient with cerebellar hemorrhage). A total of 33 patients exhibited no symptoms (Table 1); many of these were identified incidentally in studies for headache, vertigo and other conditions.[6,11,12] In particular, we have experienced four cases presenting with only mild headache or neck pain and no other clinical manifestations. In such cases, diagnosis relies exclusively on imaging findings, specifically, intramural hematoma, double lumen, stenosis, and fusiform dilatation. Sites of occurrence were predominantly located in the basilar and vertebral arteries, though a few cases were located in the internal carotid artery, middle cerebral artery, or anterior cerebral artery (ACA). Acute infarction in the ACA territory alone is especially characteristic of ACA dissection in young patients.[13–15] The culprit artery was often a branch of the vertebral artery; in several patients, for example, it was the posterior inferior cerebellar artery (PICA) alone.[12,16] It is worth noting that bilateral vertebral artery dissection was also frequently seen.

In each of the imaging modalities, we retrospectively examined the findings associated with acute and chronic phase dissection. In acute phase dissection

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Table 1 Characteristics of all patients treated at a single institute

Variable (n = 107)		(%)			(%)
Age, mean age ± SD	59.2 ± 14.9		Dissecting site	VA	58 (54)
Gender, men:women	71:36			BA	6 (6)
Onset, acute:chronic	73:34			VA–BA	16 (15)
Clinical manifestation	Infarction	63 (59)		VA–PICA	4 (4)
	Hemorrhage	13 (12)		PICA	6 (6)
	Headache	3 (3)		AICA	1 (<1)
	Vertigo	3 (3)		PCA	2 (2)
	Neck pain	1 (<1)		ACA	5 (5)
	Incidental	20 (19)		MCA	1 (<1)
	Unknown	4 (4)		ICA	8 (7)

ACA: anterior cerebral artery, AICA: anterior inferior cerebellar artery, BA: basilar artery, ICA: internal carotid artery, MCA: middle cerebral artery, PCA: posterior cerebral artery, PICA: posterior inferior cerebellar artery, VA: vertebral artery.

Table 2 Distribution of findings and modalities in acute dissection (n = 73)

No.	Double lumen	Intramural hematoma	P & S	Fusiform dilatation	Occlusion	Stenosis	Saccular aneurysm
MRA	9	3	5	7	19	24	0
T_1WI	2	26	0	0	0	2	0
3D-GdT_1WI	19	2	0	5	4	3	0
MSDE	2	14	0	0	0	1	0
CTA	1	0	3	5	7	11	0
Angiography	1	1	13	8	6	8	0

CTA: CT angiography, MRA: MR angiography (source images), MSDE: multisection motion-sensitized driven-equilibrium, P & S, pearl and string sign, T_1WI, T_1-weighted image, 3D-Gd T_1WI: three-dimensional T_1-weighted image with Gd contrast media.

(n = 73), MR angiography (MRA) source image, T_1-weighted image (T_1WI) and three-dimensional T_1WI with contrast media (3D-GdT_1WI) are likely to show double lumen and intramural hematoma. Catheter angiography is likely to detect the pearl and string sign and fusiform dilatation. MRA is likely to detect stenosis and occlusion (Table 2). In chronic phase dissection (n = 34), T_1WI and 3D-GdT_1WI are likely to detect double lumen and intramural hematoma. MRA and 3D-GdT_1WI are likely to detect stenosis and occlusion as well (Table 3). In particular, the detection of double lumen and intramural hematoma in MRA source images, T_1WI, and 3D-GdT_1WI is characteristic of arterial dissection and is therefore a key factor in diagnosis. Yet we found that cerebral arterial dissection is associated with a variety of image findings in each modality. Diagnosis of cerebral arterial dissection should accordingly be made based on multi-modal findings.

Conventional Imaging Methods and Findings Associated with Cerebral Arterial Dissection

Image analysis plays an important role in the diagnosis of cerebral arterial dissection. The most frequently used imaging methods are conventional angiography, CT, and MRI. In this section, we will discuss the findings that are commonly associated with cerebral arterial dissection in each of these imaging methods.

Angiography

Catheter cerebral angiography including the four-vessel study continues to play an important role in diagnosing cerebral arterial dissection. This method enables us not only to delineate the shapes of the arteries, but also to evaluate flow in the vessels. This method is especially useful in patients with

Table 3 Distribution of findings and modalities in chronic dissection (n = 34)

No.	Double lumen	Intramural hematoma	P & S	Fusiform dilatation	Occlusion	Stenosis	Saccular aneurysm
MRA	3	0	3	1	6	9	1
T_1WI	1	6	0	0	0	0	0
3D-GdT_1WI	11	0	0	2	4	5	0
MSDE	0	2	0	0	0	0	0
CTA	1	0	1	1	1	2	0
Angiography	0	0	0	2	0	2	0

CTA: CT angiography, MRA: MR angiography (source images), MSDE: multisection motion-sensitized driven-equilibrium, P & S, pearl and string sign, T_1WI, T_1-weighted image, 3D-Gd T_1WI: three-dimensional T_1-weighted image with Gd contrast media.

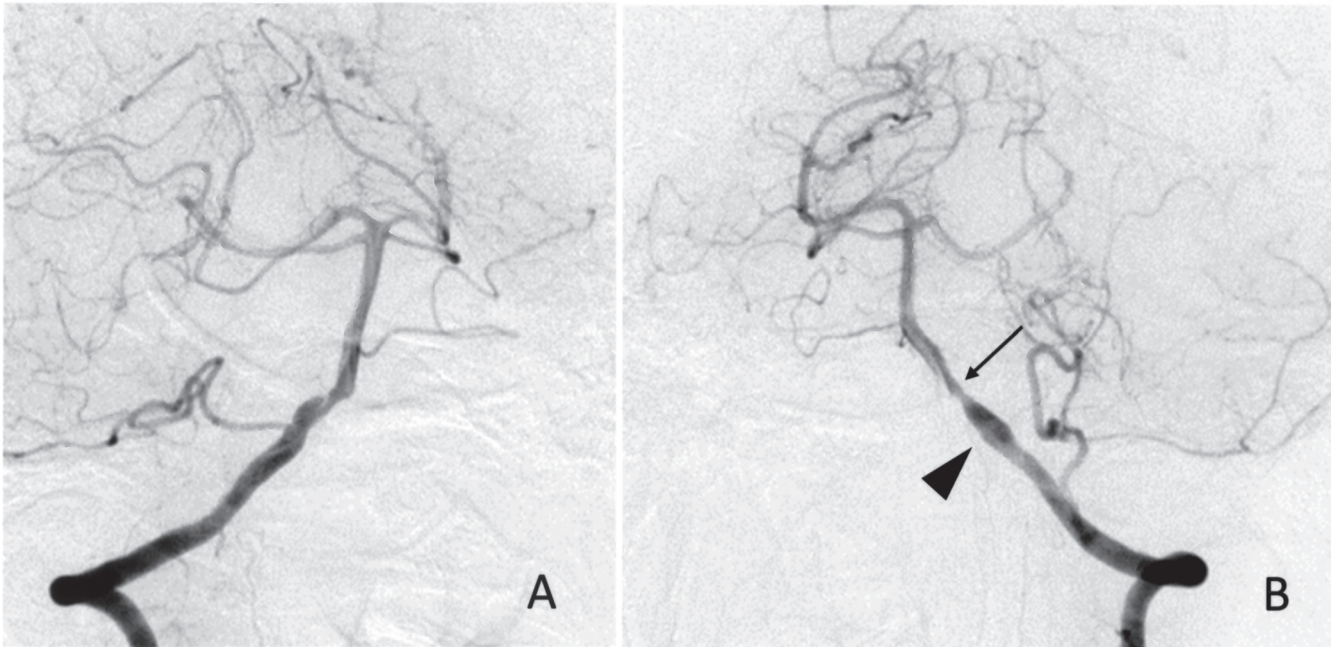

Fig. 1 53-year-old male. Bilateral vertebral artery dissection. Catheter angiography (3 days from onset) shows right vertebral artery irregularity (A), left vertebral artery proximal dilatation (B, ▼) and distal stenosis (B, →).

subarachnoid hemorrhage. "Intimal flap" and "double lumen" are the most characteristic findings associated with cerebral arterial dissection, though, in practice, these findings are visible in few cases. CT angiography and MRI are superior to conventional cerebral angiography in delineating intimal flap and double lumen.[4–6]

On the other hand, the "pearl and string sign" is a relatively specific finding that is often seen in conventional cerebral angiography. Both fusiform aneurysmal dilatation and stenosis at proximal and/or distal sites are also commonly seen. The "string sign", which consists of long-segment stenosis, has a sawtooth appearance and is also a specific finding (Figs. 1, 2).[17–20] Retention of contrast media may be seen in the pseudolumen.[21] As intramural hematoma shrinks, these findings are improved. Fusiform aneurysmal dilatation and tapered stenosis are sometimes present but are not always indicative of cerebral arterial dissection (Fig. 3).

As rapid injection of the contrast media for this imaging modality may cause deterioration of the arterial dissection, it is crucial to monitor and control injection rate and volume.

CT angiography (CTA)

Multidetector CT angiography (64–320 rows) is a very rapid, precise, and broad-scanning form of

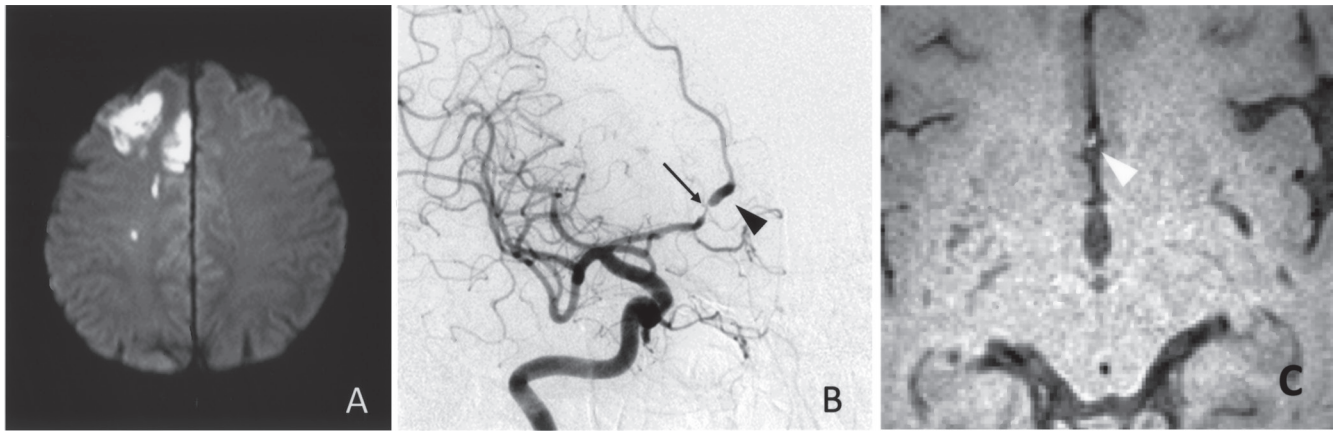

Fig. 2 54-year-old male. Right anterior cerebral artery territory infarction due to anterior cerebral artery dissection. Diffusion weighted image (2 days from onset) reveals anterior cerebral artery territory acute infarction (A). Catheter angiography (16 days) reveals proximal stenosis (→) and distal dilatation (▼), also known as the "pearl and string sign", in right anterior cerebral artery (B). Three-dimensional multisection motion sensitized driven equilibrium (12 days) shows high intensity crescent (▽).

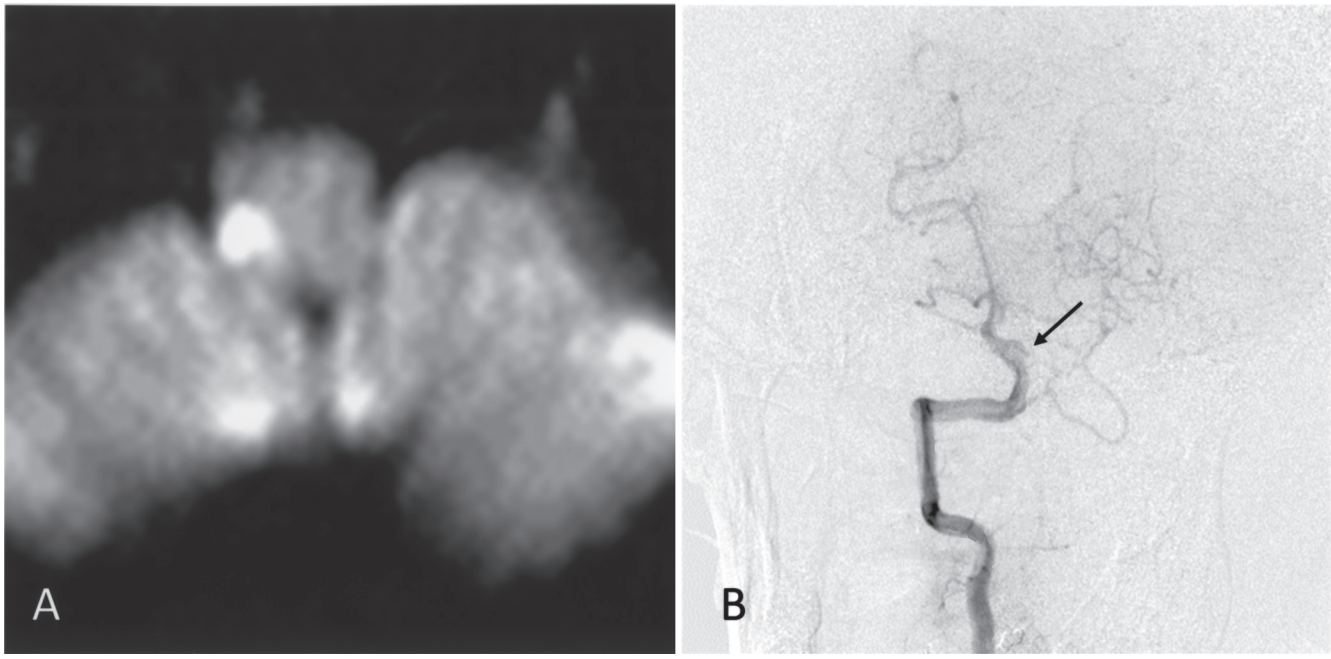

Fig. 3 44-year-old male. Wallenberg syndrome due to right vertebral artery dissection. Diffusion weighted image (0 days from onset) reveals right lateral medullary infarct (A). Catheter angiography (0 days) reveals fusiformic dilatation in right vertebral artery (→) (B).

angiography that requires an intravenous bolus injection (3.5–4.5 ml/s). CTA can be performed more safely and more rapidly than catheter angiography while enabling the detection of many of the same signs. Like conventional CT, multidetector CT can now be used for 4D angiography. It also enables the detection of arterial stenosis occlusion and fusiformic dilatation in arterial dissection patients, as like catheter cerebral angiography (Fig. 4). It may also permit the identification of the double lumen and intimal flap signs.[17,22] Finally, CTA source imaging may enable us to observe pseudolumen through its visualization of wall thickness.[23] CTA is thus superior to MRA in the detection of arterial dissection.[20,24] In CTA, however, arterial dissection can resemble atherosclerosis, so that we must take particular care in distinguishing the two conditions.

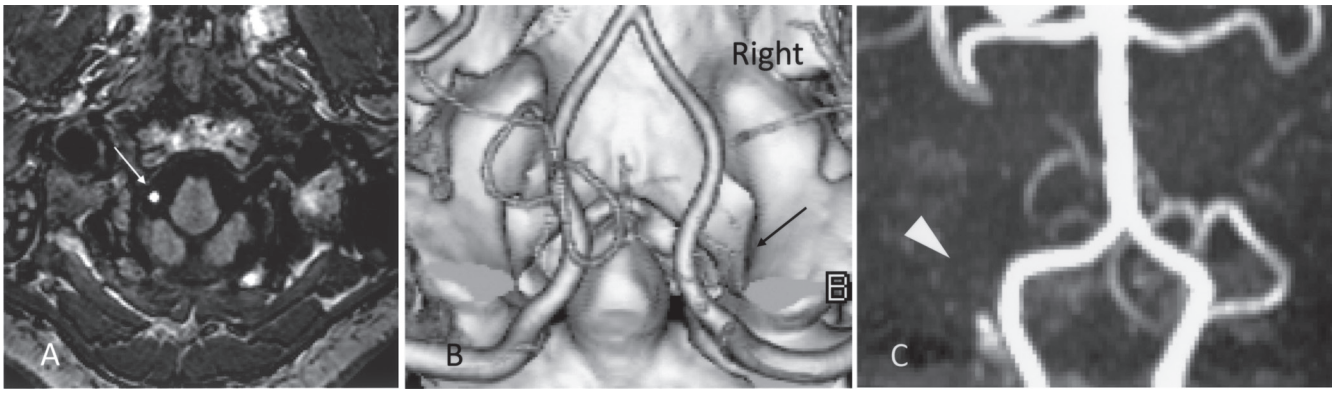

Fig. 4 54-year-old female. Right posterior inferior cerebellar artery dissection. Multisection motion sensitized driven equilibrium (1 month from onset) reveals high-intensity intramural hematoma in right posterior inferior cerebellar artery (A, →). Volume rendering image of CT angiography (1 month) reveals right posterior inferior cerebellar artery irregularity in comparison with left posterior inferior cerebellar artery (B, →). Magnetic resonance angiography (1 month) shows no delineation of right posterior inferior cerebellar artery (C, ▽).

MRI

MRI plays a most important role in diagnosing acute infarction and cerebral arterial dissection through non-invasive means. MRA can delineate arterial shape, while three-dimensional volume imaging can delineate intramural hematoma and double lumen related to arterial dissection. Intramural hematoma appears as a region of high intensity on T_1WI. Thus, this imaging procedure should be performed with the thinnest possible slices (<4 mm axial images). The most up-to-date high magnetic field MRI units such as the 3-Tesla units can yield images with particularly high resolution.

MRA: Generally speaking, MRA can be performed easily and is useful in screening for aneurysm and arterial stenosis.[25] MRA can delineate the shapes of vessels without gadolinium contrast media as well as conventional catheter cerebral angiography can. High magnetic field MRI units, i.e., 3.0-Tesla or stronger units are preferable as they provide higher resolution MRA images. Like conventional catheter cerebral angiography, MRA can reveal the "pearl and string sign", "string sign", fusiform aneurysmal dilatation, and so on. Double lumens and intimal flaps may be seen through careful observation of source images in MRA (Fig. 5).[17,25] Cautious examination of the images is necessary, however, as a true double lumen or intimal flap may be mistaken for a flow artifact associated with the time-of-flight method.

3D-T_1WI: Intramural hematoma is frequently observed in dissecting arteries, where it appears as a typical change in signal intensity related to the paramagnetic effects of the products of hemoglobin breakdown. In the early stage and again in the chronic stage, the hematoma is usually isointense to surrounding structures, whereas between 7 days and 2 months, it is almost invariably bright in T_1WI.[17,26] The ability to detect intramural hematoma through T_1WI is important (Fig. 6). Yet, conventional T_1WI uses slices that are too thick (usually 6 mm) to detect intramural hematoma. Thin-slice three-dimensional T_1WI (3D-T_1WI) is therefore useful in detecting intramural hematoma and intimal flap.[11,17,19,26,27] These methods yield multi-planar reconstruction (MPR) images, volume rendering, and the ability to observe the vessels from all angles. 3D-T_1WI may be useful in detecting small intramural hematoma.

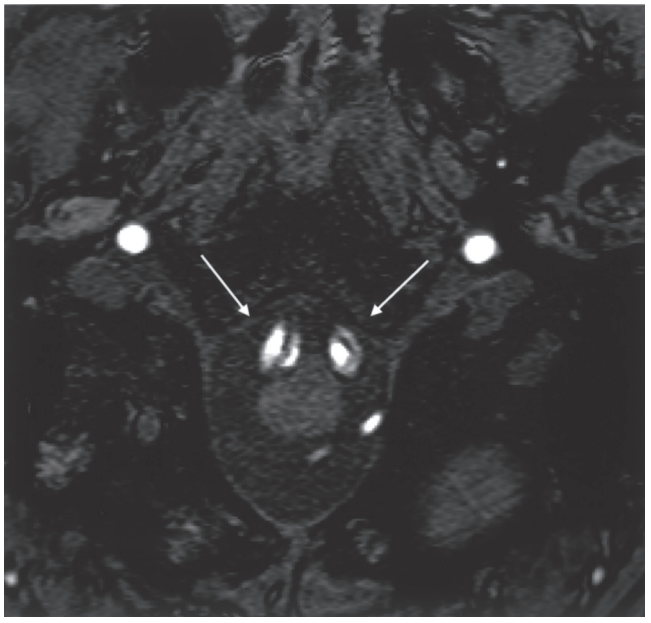

Fig. 5 53-year-old male. Bilateral vertebral artery dissection. Magnetic resonance angiography source images (3 days from onset) reveal double lumen in bilateral vertebral artery (→).

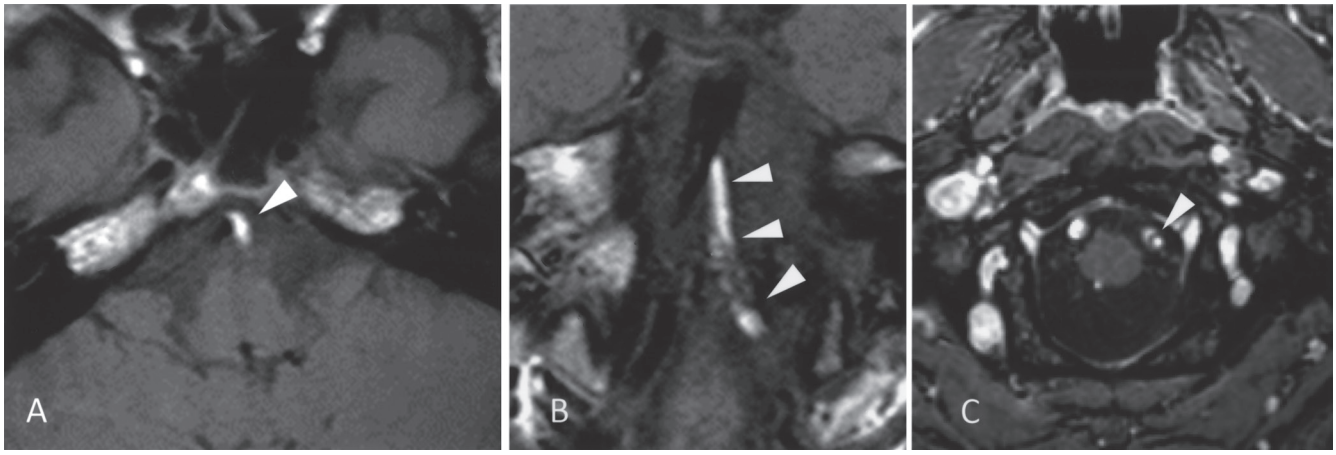

Fig. 6 37-year-old female. Left vertebral artery to basilar artery dissection. Axial and coronal T_1-weighted images (3 days from onset) reveal high-intensity intramural hematoma (A, B, ▽). Double lumen in left vertebral artery is revealed by Gd-T_1TFE (C, ▽).

Moreover, 3D-T_1WI axial imaging with gadolinium contrast media (3D-CE-T_1WI) also enables us to delineate the double lumen and the intimal flap. It may be difficult, however, to distinguish small intramural hematoma from lipid-rich atherosclerotic plaque. In such cases, it is important to monitor changes of the intramural hematoma on follow-up MRI. We should be especially suspicious of brain stem infarction in young adults.

Susceptibility-weighted imaging (SWI): SWI has been used in the detection of intracranial hemorrhage and in the measurement of oxygen conditions in the venous blood and thrombus. SWI provides us with higher resolution images than T_2-weighted gradient echo imaging (T_2*WI). Thus, SWI is useful for visualizing intracranial microhemorrhage. In cerebral arterial dissection, a crescent dark rim may be indicative of intramural hematoma.[27]

Basiparallel anatomic scanning (BPAS): Basiparallel anatomic scanning (BPAS) is useful as a supplementary imaging modality in diagnosing cerebral arterial dissection. BPAS is a heavily T_2*WI method that can be performed easily. It provides coronal images of the vertebrobasilar artery with thick slices (20 mm), allowing us to observe the outer shape of the vertebrobasilar artery. This enables us to detect fusiform aneurysmal dilatation due to arterial dissection, which cannot be detected with MRA alone. It is difficult to make a final diagnosis of arterial dissection with BPAS alone, but it is a useful method for screening of arterial dissection (Fig. 7).[28] In addition, this modality also allows us to distinguish arterial stenosis or occlusion due to dissection from hypoplasty of the vertebral arteries, a distinction that cannot be made with such accuracy based on MRA alone.[29]

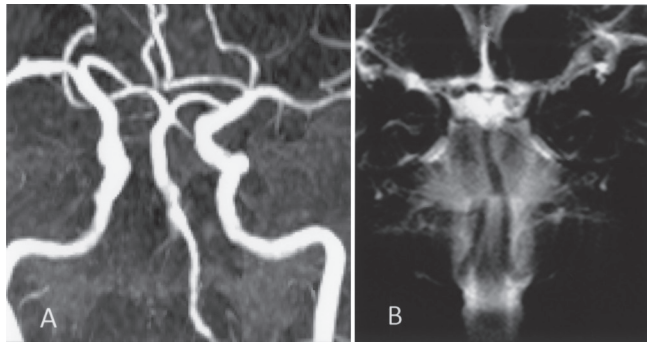

Fig. 7 53-year-old female. Right vertebral artery dissection. Magnetic resonance angiography (2 days from onset) shows occlusion of right vertebral artery (A). Basiparallel anatomic scanning reveals outer shape of right vertebral artery is normal (B). These findings indicate vertebral artery occlusion because of arterial dissection.

New MRI Method; Multisection Motion Sensitized Driven Equilibrium (MSDE)

MSDE is a novel MRI technique similar to diffusion weighted imaging (DWI). As a preparative sequence, this method begins with a diffusion pulse with a very low b-value consisting of three nonselective RF pulses with flip angles of 90–180–90° and symmetric gradients around the 180° pulse. MDSE causes phase dispersion of blood spin using a magnetic field gradient and suppresses the blood flow signal, and 3D T_1 or T_2*WI can thus be acquired.[30] MSDE is used to detect brain tumors and to delineate carotid arterial atherosclerotic plaque, thin cranial nerves, and similar structures.[31–34] Intramural hematomas appear as regions of hyperintensity in this modality because MSDE can cancel any flow signal without an in-flow

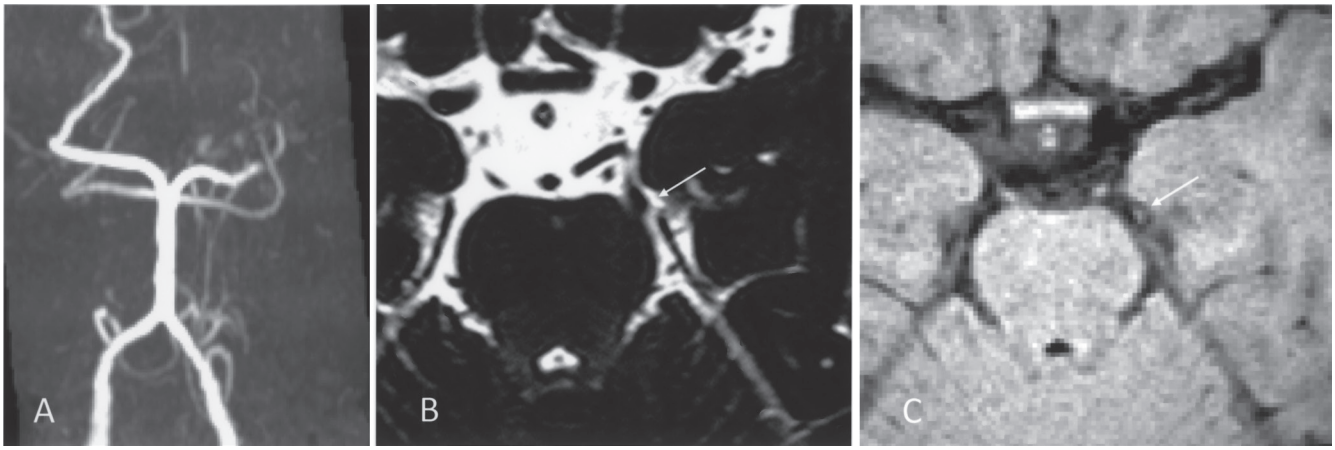

Fig. 8 70-year-old female. Left posterior cerebral artery dissection Magnetic resonance angiography (10 days from onset) shows occlusion in left posterior cerebral artery P2 segment (A). Magnetic resonance cisternography shows normal outer shape of proximal left posterior cerebral artery P2 segment (B, →). Thin slice multisection motion sensitized driven equilibrium (0.8 mm) reveals small crescent isointensity intramural hematoma in the proximal left posterior cerebral artery P2 segment (C, →).

effect. Because thin-slice images (0.8 mm) can be obtained, MSDE enables us to delineate small intramural hematomas as high-intensity lesions in small arteries such as the PICA, AICA, and distal ACA (Figs. 2c, 8). In practice, however, it is difficult to distinguish intramural hematomas from atherosclerotic plaque.

Recommendation of Diagnostic Procedure for Cerebral Arterial Dissection

Cerebral arterial dissection is associated with a variety of imaging findings in the various imaging modalities that are used in its diagnosis. It would be helpful to identify a useful comprehensive method for the diagnosis of cerebral arterial dissection based on the results of multiple modalities. In summary, we will discuss the diagnostic procedure for cerebral arterial dissection through our analysis of acute and chronic dissection cases (Fig. 9).

Diagnosis of acute dissection

Dissection with hemorrhage: Hemorrhagic dissection results in subarachnoid and/or intracranial hemorrhage. For patients with suspected hemorrhage due to acute arterial dissection, it is necessary to perform CTA to confirm the presence of a double lumen, the pearl and string sign, and fusiform aneurysm and to rule out ruptured saccular aneurysm. Afterward, catheter angiography is necessary to confirm these findings and to perform embolization.

Dissection with ischemic lesions: A combination of BPAS and MRA is useful in screening for arterial dissection in the posterior fossa, because a discrepancy in diameter between the BPAS and MRA results indicates a strong possibility of arterial dissection. Arterial dissection should also be considered when acute infarction is observed in the posterior fossa or in the ACA territory only, particularly in young patients. MRI is a very important modality in the definitive diagnosis of cerebral arterial dissection. Many arteries, including small branch arteries such as the peripheral PICA, AICA, SCA, PCA, and ACA, must be examined for double lumen, intramural hematoma, fusiform dilatation, and the pearl and string sign, and MRA source imaging, T_1WI, 3D-GdT_1WI, and other forms of MRI make this feasible through their sensitivity to arterial changes. Afterward, CTA should be performed to confirm arterial stenosis and occlusion. MRI and CTA can be performed as follow-up studies of cerebral arterial dissection because they are minimally invasive. In addition, catheter angiography should be performed if MRA and CTA cannot detect arterial dissection findings (Fig. 9, Table 2). Cerebral arterial dissection can cause mild headache and neck pain, and these can even be the only symptoms, as we have seen in four cases lacking any other clinical manifestations. As MRI alone is not capable of showing intramural hematoma, double lumen, stenosis, and fusiform dilatation, T_1WI and MRA source images must be examined carefully in such cases.

Chronic dissection

MRI is very important in the diagnosis of chronic cerebral arterial dissection, which includes asymptomatic and incidental cases. One MRI finding frequently seen in chronic dissection is brain stem

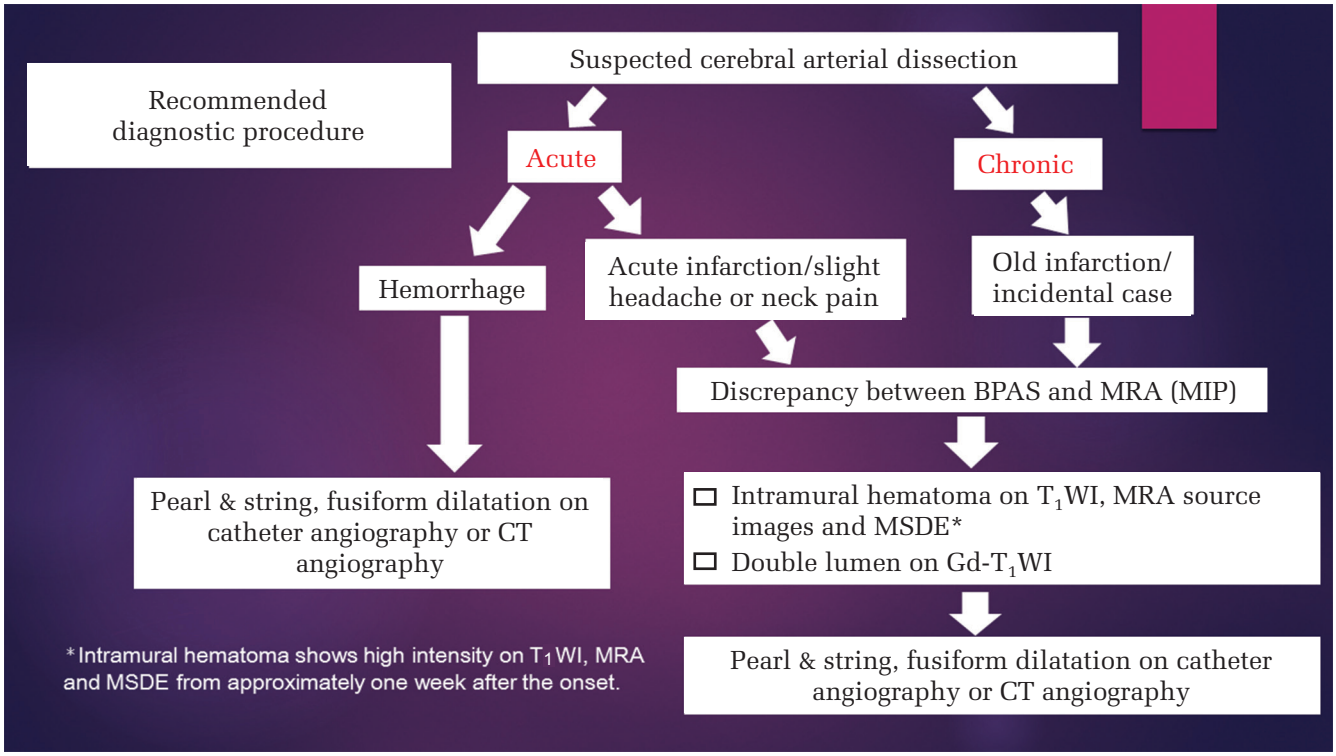

Fig. 9 Recommended diagnostic procedure

and cerebellar infarction. BPAS and MRA are also useful in screening chronic cerebral arterial dissection. When a discrepancy between BPAS and MRA findings is noted, cerebral arterial dissection should be suspected. Next, T_1WI, 3D-GdT_1WI, MRI source images, MSDE, and similar modalities should be used to test for double lumen and intramural hematoma. Any of these findings should lead to a suspicion of cerebral arterial dissection (Fig. 9, Table 3). MRI is also useful in follow-up studies.

Limitations of image-based diagnosis

Cerebral arterial dissection should be diagnosed based on a combination of CT and MRI. In many patients, whose clinical manifestations are suspicious of cerebral arterial dissection, these modalities are likely to reveal intramural hematoma and double lumen. Especially among elderly patients, however, it is difficult to definitively distinguish intramural hematoma from atherosclerotic plaque. We should be mindful of the limitations of each modality when diagnosing cerebral arterial dissection (Table 4). Furthermore, in cases of true arterial dissection, the imaging findings will change over time. We should carefully observe the changes in these findings through follow-up studies.

Table 4 Major limitations of imaging modalities

Modality	Limitation
MRA	Mistaking double lumen for flow artifact.
T_1WI	Mistaking intramural hematoma for in-flow effect.
3D-GdT_1WI	Mistaking double lumen for flow artifact.
MSDE	Data often insufficient to distinguish intramural hematoma from atherosclerotic plaque.
CTA	Mistaking double lumen for flow artifact.
Angiography	Data insufficient to depict intramural hematoma.

Conclusion

Cerebral arterial dissection is a serious disease characterized by acute onset. Rapid and precise diagnosis enables appropriate treatment and good prognosis. As several imaging methods (conventional angiography, CT, and MRI) can be used in diagnosis, and as these modalities show different findings in connection with cerebral arterial dissection, it is essential to select the most appropriate imaging

method and to understand the findings associated with each method.

Conflicts of Interest

This study was not affected by any conflicts of interest pertaining to the authors or our institute.

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