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An Overview About Intracranial Dissection: Pathogenesis and Imaging

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Abstract: Intracranial artery dissections (IAD) are uncommon entities associated with high rates of morbidity and mortality. Certain ethnic groups and patients with underlying connective tissue disorders may be at a higher risk of developing IAD, but these relationships are unclear due to the condition's rarity. Patients often present with a prodromal headache followed by subarachnoid hemorrhage (SAH) or ischemic stroke. Imaging findings are critical to establishing the diagnosis, as the lesions have a myriad of presentations based on the severity, location, and timing of the dissection. Lesions that present with ischemia are at high risk for future ischemia but low risk of future hemorrhage, whereas lesions, which present with hemorrhage have a high rate of re-bleeding if left untreated. There are no evidence-based guidelines for medical or surgical management. Several endovascular and surgical techniques have been used to prevent or treat hemorrhage by ligating the parent artery or reconstructing the vessel wall. Outcomes are generally poorer in patients with IAD than cervical artery dissection, particularly in those who suffer SAH.

Keywords: *Dissection, intracranial, stroke, vessel wall imaging*

1. Introduction

While much is known about the pathogenesis, presentation, and prognosis of cervical artery dissections, intracranial artery dissections (IAD) are much rarer and more poorly characterized. There are relatively few studies of IAD in the literature, but they seem to have a predilection for young adults and arteries of the posterior circulation [1], [2]. There are numerous mechanisms by which IAD can arise, and each has a different clinical presentation and imaging findings. Patients most often present with a nonspecific headache followed by ischemic stroke or subarachnoid hemorrhage (SAH). The heterogeneity of this rare condition precludes standardized diagnostic criteria and evidence-based treatment guidelines. In this comprehensive review, the epidemiology, pathogenesis, clinical and radiographic features, and natural history of IAD are discussed.

Epidemiology:

There are no population-based studies to date that estimate the incidence of IAD. However, it is uncommon than spontaneous extracranial carotid and vertebral artery dissections, which have a combined incidence of 3.6 to 4.4 per 100,000 people per year [3-7]. While IAD can occur in both pediatric and adult populations, there seems to be a predilection for young adults. The mean age at the time of presentation is 50 years [1].

In adults, men and women are equally affected. In children, there seems to be a preponderance to affect males that is not explained by trauma [7]. Most information about IAD is extrapolated from series of carotid and vertebral artery dissections that include both intracranial and extracranial segments. Studies from different geographic regions with substantially different ethnic populations report different ratios of intracranial to extracranial dissection.

In two studies with exclusively East Asian patients, 67% and 78% of cervico cerebral dissections were intracranial [8,9]. A study from Mexico reported that 27 of their 110 patients with vertebral artery dissections were intracranial [10]. In a Portuguese population study, 12% of dissections were intracranial [11]. Of 169 vertebral artery dissections in a series published by two Western European institutions, 11% were intracranial [12]. These seemingly large differences between regions could suggest an underlying risk for IAD based on ethnicity. Notably, each of these studies had significantly different recruitment methods and inclusion criteria. For instance, case series published by neurology departments reported more patients who presented with ischemic stroke, whereas neurosurgery studies included more patients with SAH. In light of these methodological differences, these studies must be interpreted with caution. (12)

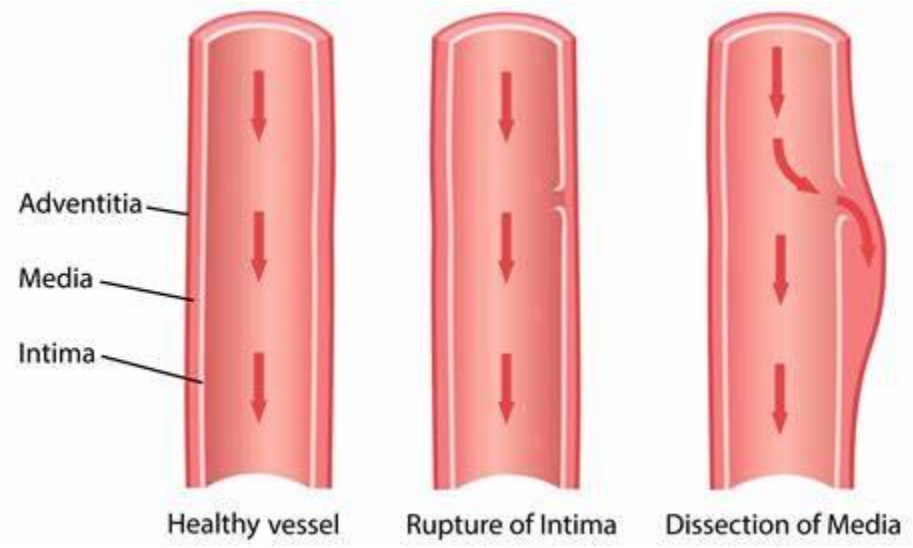

Figure (1): Pathogenesis of vessel Dissection (13)

Risk factors and predisposing conditions:

Risk factors for intracranial artery dissections are unknown. No comparisons exist between putative risk factors in patients with intracranial artery dissection and healthy controls. In the few studies that included both patients with cervical artery dissection and those with intracranial artery dissection, distribution of vascular risk factors did not differ between the two groups (13) except for one study (14) showing a higher prevalence of hypertension in patients with intracranial artery dissection. However, this finding (13) might be accounted for by the older age of patients with intracranial artery dissection than control participants in that study (mean age 48 years vs 37 years).

Whether cervicocerebral trauma is a risk factor for intracranial artery dissection, as it is for cervical artery dissection, (15) is unclear. In two studies (13) that compared patients with cervical artery dissection and patients with intracranial artery dissection, a history of minor trauma was more often present in patients with cervical artery dissection, both in children and adults. In our experience, sudden physical movements that lead to a sudden stretch of the arteries are sometimes reported before the event, but this association has not been systematically analysed in large patient series.

Some instances of intracranial artery dissection in children, have been associated with intracranial or systemic infections. Differences in prevalence and characteristics of intracranial artery dissections between ethnic origins, and the more frequent occurrence of intracranial artery dissection in children than in adults, suggest

that genetic risk factors could contribute to the occurrence of intracranial artery dissection. However, genetic contribution to intracranial artery dissections has so far not been explored. Exceptionally, intracranial artery dissection might be a complication of rare monogenic disorders of connective tissue, such as Loeys-Dietz syndrome. (16,17) Whether carotid and vertebral artery dissections noted in patients with vascular Ehlers-Danlos syndrome include intracranial artery dissections is not detailed in reported large series. (18)

Isolated cases of suspected intracranial artery dissections have been noted in patients with Marfan's syndrome.(18)Patients with fibromuscular dysplasia (a nonatherosclerotic, non-inflammatory vascular disease that mainly affects the renal and cervical arteries) have an increased risk of cervical artery dissection and intracranial aneurysms; whether these patients also have an increased prevalence of intracranial artery dissection is unknown.(19) Only isolated instances of intracranial artery dissection and fibromuscular dysplasia have been reported (20) and patients with fibromuscular dysplasia were excluded from many reported series of patients with intracranial artery dissection. Overlap between intracranial artery dissection and segmental arterial mediolysis (a rare arterial disease that presents with life threatening hemorrhages through ruptured aneurysms in the abdominal cavity), the retroperitoneum, and more seldom the base of the brain, is unclear. (20)

Mechanisms and pathological features:

Little is known about the pathophysiology of intracranial artery dissection. Although available neuropathological specimens have generally shown a disruption of the internal elastic lamina and the media(21,22) whether direct bleeding of vasa vasorum (small blood vessels in the wall of larger blood vessels) in the arterial wall can be the initial event is unclear.(23) Vasa vasorum are not always seen in intracranial arteries and seem to predominate in the tunica adventitia and proximal intracranial arteries.(24) In a study(21) where tissue samples were obtained by surgery or autopsy at different timepoints after symptom onset, the intramural hemorrhage was replaced by granulation tissue after 14 days from onset, followed by compensatory intimal thickening around the pseudo lumen.

In samples obtained after more than 30 days from symptom onset, neo vascularization in the thickened intima was reported, leading to chronic fusiform aneurysm formation, possibly aided by repetitive intramural hemorrhage with the rupture of fragile neovessels.(25) Different patterns of intimal injury in intracranial artery dissection have been reported. A mural hematoma might be caused by one entrance in the pseudo lumen (so-called entry-only lesions) or an entrance and an exit in the pseudo lumen (so-called entry-exit lesions).

Entry only lesions can have a higher occurrence of subarachnoid hemorrhage than entry-exit lesions. (26) The pathophysiological overlap of intracranial artery dissection with giant fusiform aneurysms and blood blister-like aneurysms is controversial, but they should probably be regarded as distinct entities. (27-30) Mycotic or oncological giant fusiform aneurysms are non-dissecting and are caused by the release of proteases by bacteria or tumor cells that break down the vessel wall. Blood blister-like aneurysms located at non-branching sites of intracranial arteries are caused by a degeneration of the internal elastic lamina and media without associated arterial dissection (no mural hematoma or double lumen on pathological examination).

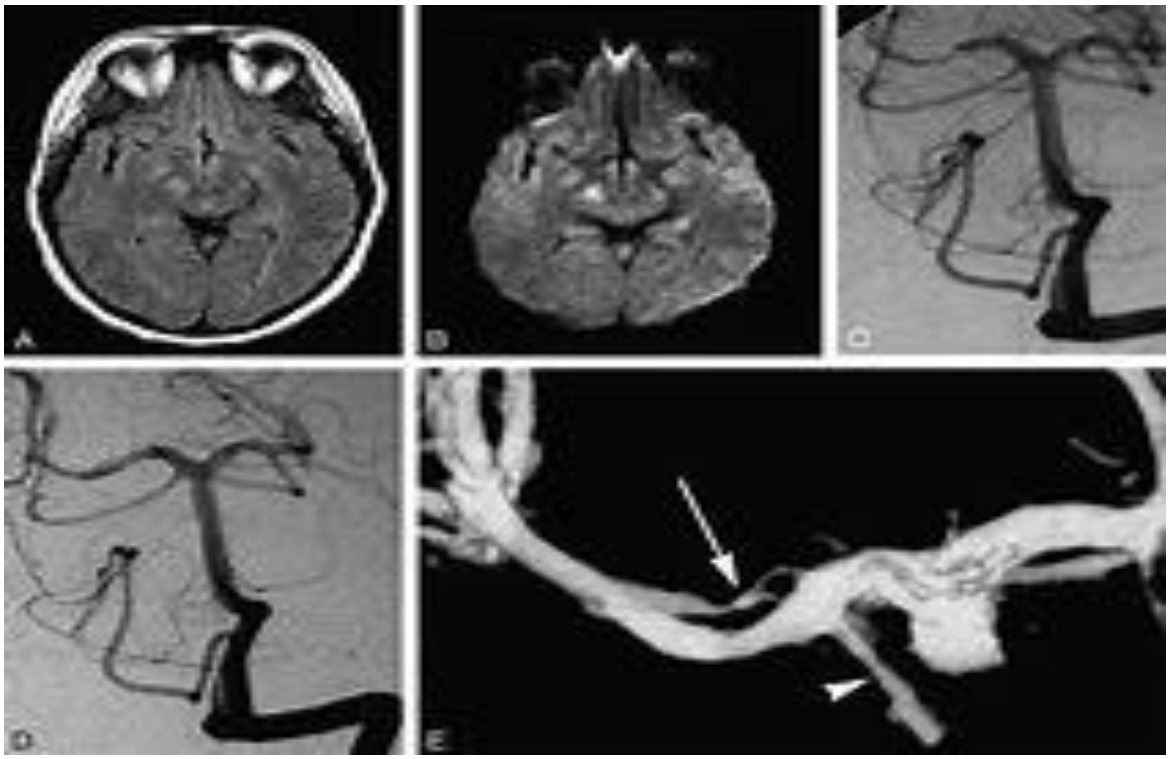

Figure (2) *A*, Axial FLAIR image demonstrating hyperintensity in the right cerebral peduncle.
B, Axial diffusion-weighted image (DWI) image showing corresponding abnormal increased signal intensity. There was respective low signal intensity on the apparent diffusion coefficient map (not shown).
C, DSA, left vertebral artery injection, transfacial view, demonstrates a smoothly tapered focal, near-occlusive narrowing of the proximal right PCA.
D, DSA, left vertebral artery injection, transfacial view. One-year follow-up demonstrating a "double lumen" sign of the right P2 segment.
E, 3D reconstruction of left vertebral angiogram rotational DSA which shows 2 patent lumens (double lumen sign) distal to the posterior communicating artery (*arrowhead*). (31)

Imaging findings:
Radiological diagnostic criteria for intracranial dissection, which include the following:
• fusiform or irregular aneurysmal dilation at a non-branching site; long, filiform, irregular stenosis with one or more of the following: double lumen, intramural hematoma, intimal flap, rapid morphologic change on repeat imaging, focal stenosis and dilation ("string and pearl sign");
or: • arterial occlusion with recanalization at a non-branching site with fusiform or irregular aneurysmal dilation [1]. These criteria are based primarily on conventional angiography, CTA, or MRA imaging findings. Ultimately, some of these imaging appearances may be non-specific on conventional imaging as there is a broad differential diagnosis for focal aneurysmal dilatations, stenosis and occlusions, including post-stenotic dilatation in the setting of atheromatous disease.
While subintimal dissections are readily recognizable on luminal imaging from the appearance of the true and false lumen, sub-adventitial dissections may be more difficult to detect and diagnose.

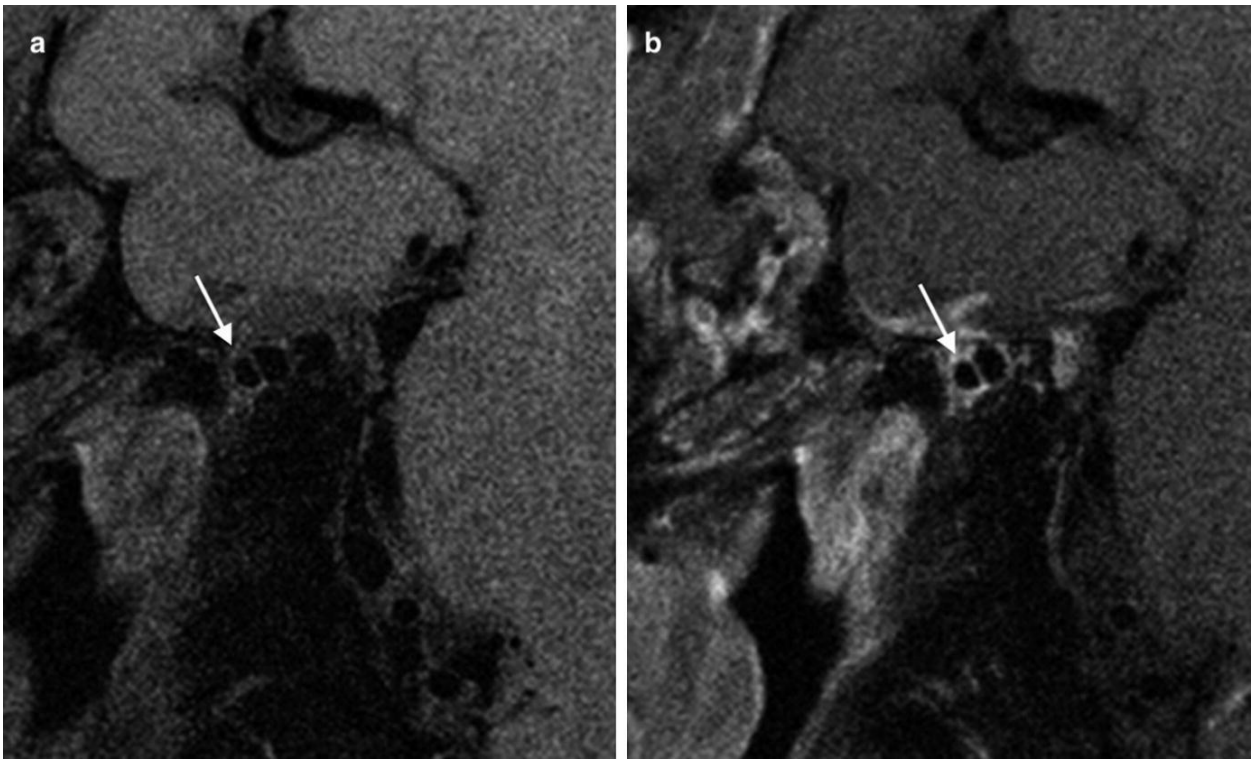

Figure (3) Chronic arterial dissection of the horizontal petrous segment of the carotid artery. Non-contrast (**a**) and contrast-enhanced (**b**) VWMR images through the short axis of the artery demonstrate enhancement of the vessel wall and flap separating the false lumen (arrow) from the true lumen (32)

Imaging Dissections Causing Ischemia:

Dissections resulting in ischemia typically manifest with a long filiform irregular stenosis with double lumen, intramural hematoma or intimal flap [33]. T1 weighted HR-VWI has been shown to be helpful in the detection of high signal intramural hematoma associated with subadventitial dissections [34]. HR-VWI has also been shown to be sensitive in detecting characteristics such as intimal flap and double lumen, findings that are specific to dissection.

Vessel wall contrast enhancement is sometimes seen in intracranial dissection and is thought to be secondary to a combination of slow blood flow in the false lumen, inflammation and enhancement of the vasa vasorum. While both dissections and atherosclerotic plaque with intraplaque hemorrhage can demonstrate luminal narrowing with T1 hyperintensity, and eccentric vessel wall enhancement, other findings including an intimal flap, double lumen, or clear crescentic false lumen should indicate dissection. T2W VWI has also been shown to be useful in characterizing intracranial dissections as the intramural hematoma can demonstrate hyperintensity or hypointensity depending on the age of the dissection. High-resolution susceptibility weighted imaging has a high sensitivity for detecting intramural hematoma.

Management Dissections and Ischemia:

There are currently no evidence-based guidelines for the treatment of IAD due to the condition's rarity. Medical management includes antithrombotic or antiplatelet therapy for the prevention of thromboembolic stroke. Several randomized-controlled trials and retrospective studies have found no difference between antiplatelets and anticoagulants for stroke prevention in patients with cervical artery dissection and these data have at times been extrapolated to the intracranial vasculature [35–38].

No equivalent studies have been performed in patients with IAD. Similarly, intravenous thrombolysis therapy has been deemed a safe treatment in patients with cervical arterial dissection who present with ischemic stroke and is thus thought to be safe in patients with intracranial dissection, although supportive data are limited to case reports [39].

In patients with IAD resulting in large vessel occlusion, there is no contraindication to endovascular recanalization. However, these lesions will sometimes behave similar to active atheromatous plaques with short-term re-occlusion of the parent artery. Some authors have reported a low threshold to perform stenting in these patients. There will be a subset of patients who suffer from recurrent strokes despite medical therapy. In these patient's itis reasonable to consider options such as stent reconstruction with self-expanding stents and/or surgical bypass. Such strategies have been reported with a fair amount of success in prior case reports and small case series.

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