Benign arterial dissections of the posterior circulation

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Four young adults with spontaneous dissection of the vertebrobasilar system are reported. Clinically, two patients presented with subarachnoid hemorrhage and two with brain-stem ischemia. In two cases of ruptured arterial dissection of the posterior cerebral artery, angiography demonstrated fusiform and "sausage-like" dilatation of the involved vessel. In two cases of occlusive dissection of the basilar artery, angiography revealed the typical "string sign." All four patients were treated conservatively: three survive in good clinical condition and one remains disabled. Follow-up angiograms showed spontaneous healing of the lesion with return to an almost normal arterial configuration in two cases; residual narrowing corresponding to the dissection was the most notable finding in the other two. It is recommended that, in a subset of neurologically stable patients, angiographic monitoring is undertaken to assess the tendency for spontaneous repair before surgical intervention is planned.

KEY WORDS  •  aneurysm, dissecting • ischemia • subarachnoid hemorrhage  • posterior circulation  • vertebrobasilar system

While extracranial carotid dissections are characterized by a relatively benign clinical course and a good rate of healing,\textsuperscript{6,17} the natural history of intracranial dissections seems to be much more severe.\textsuperscript{1,2,4,5,11,12,15,16,19,21} In particular, survival and spontaneous cure have been reported very rarely.\textsuperscript{10} We describe four patients suffering from dissections of the vertebrobasilar system. The clinical course in these patients was benign and angiographic monitoring demonstrated spontaneous healing or improvement of the lesion.

Case Reports

Case 1

This 33-year-old previously healthy man developed sudden headache and diplopia. Neurological examination during his initial evaluation revealed a sixth nerve paresis with horizontal nystagmus. Lumbar puncture yielded normal cerebrospinal fluid (CSF). A computerized tomography (CT) scan was normal. Four-vessel angiography showed an almost complete tapering occlusion ("string sign") of the midportion of the basilar artery (Fig. 1 left). The patient’s symptoms gradually abated and he was discharged on antiplatelet medication. During the following 4 years, his condition remained stable, and a recent vertebral angiogram demonstrated that the occlusion had resolved and that the basilar artery was of normal caliber (Fig. 1 right).

Case 2

This 38-year-old woman suffered the sudden onset of headache and vomiting followed by loss of consciousness. The CSF was bloody, and a CT scan demonstrated blood in the basal cisterns and fourth ventricle. Her condition gradually cleared and four-vessel angiography 4 days after the hemorrhage showed a "sausage-like" swelling of the right posterior cerebral hemisphere.

\textbf{FIG. 1.} Case 1. Vertebral angiograms, anteroposterior view. \textit{Left:} Admission study showing an almost complete tapering occlusion ("string sign," \textit{arrow}) of the midportion of the basilar artery. \textit{Right:} Angiogram obtained 4 years later demonstrating normal caliber of the basilar artery.
artery (PCA) with diffuse narrowing of the basilar artery and the left PCA (Fig. 2 left). Repeat studies were performed 2 weeks and 1 month later. On the most recent angiograms the arterial caliber in both the dilated and narrowed segments had returned to normal; only a minor luminal irregularity of the proximal right PCA was present (Fig. 2 right). On follow-up examination 1 year after the hemorrhage, the patient remains well.

Case 3

This 44-year-old woman suffered the sudden onset of posterior neck pain and vomiting. The CSF was bloody, and a CT scan demonstrated diffuse subarachnoid and ventricular bleeding; an area of punctate hyperdensity was noted in the left peduncular region (Fig. 3). Four-vessel angiography revealed a fusiform aneurysm of the left PCA (Fig. 4 left). The patient's condition rapidly cleared and she was discharged home. Repeat vertebral angiogram 2 months later revealed diffuse narrowing of the left PCA (Fig. 4 right), and a CT scan documented a left occipital infarction. On follow-up examination 6 months later, the patient remains well.

Case 4

This 33-year-old man was well until he began to experience a dull headache. Two days later, during a go-cart race, he suffered the abrupt onset of severe headache and vomiting followed by loss of consciousness. On admission, he was comatose; he expired spontaneously and responded nonpurposely on the right side. A left hemiparesis and divergent strabismus with an enlarged left pupil were noted. The neck was supple. The CSF was normal and no abnormality was detected on the CT scans. Carotid angiography was unremarkable; vertebral angiograms demonstrated a tapering occlusion of the distal basilar artery (Fig. 5 left). On the following days the patient's condition gradually cleared: 3 weeks after the stroke he had diplopia, left-sided weakness, and a fluctuating level of consciousness. His mental status had returned to normal by the 5th week. Vertebral angiography 3 weeks after the onset of symptoms documented a residual stenosis of the distal basilar artery and segmental nodular narrowing of the PCA's with occlusion of the distal branches on the right side (Fig. 5 center and right). A CT scan showed a right occipital infarction. Eight years later, the patient was re-examined and found to have residual left hemiparesis and diplopia.
Discussion

Clinical Course

Most intracranial dissections present in young adults with ischemic stroke, and a minority present with subarachnoid hemorrhage (SAH) occurring preferentially in the vertebrobasilar system. This different clinical behavior depends partly on the location of the dissection (subintimal or subadventitial). Although the natural history of the disease is far from being understood, the frequently reported fatal outcome does represent only a part of the clinical spectrum; a benign course with minor neurological signs probably occurs more frequently than suggested in the literature. Friedman and Drake first recognized that the natural history of a bland dissection without a neurological deficit might be benign. Their Case 5 is the only recorded instance of the spontaneous resolution of a fusiform vertebral dissecting aneurysm proven by angiography.

Yamaura compared his cases of saccular and dissecting vertebral aneurysms and found that their early clinical course was similar. Most of them (75% and 81%, respectively) presented with SAH, and rebleeding occurred in 24% in both groups. In dissecting aneurysms, rebleeding occurred at a mean interval of 10.8 days. The remaining patients were stable until operation, which was always performed after 21 days.

If the tendency toward spontaneous healing found in our cases is recognized more frequently, the biological behavior of intracranial dissections may well be more favorable than expected. Spontaneous cure of intracranial carotid artery dissections has been observed by chance at autopsy in some infants who had suffered a remote stroke. In two other series, a florid reparative attempt was noted in the false lumen with proliferation of fibrous tissue and formation of a new elastic lamina and smooth-muscle fibers. In one patient with basilar artery dissection who died after aneurysm wrapping, partial healing characterized by florid myofibroblast proliferation in the media was found at autopsy 39 days after the clinical onset.

To date, no case of local recurrent intracranial dissection has been reported. In our Cases 2 and 4, an interval of 5 and 8 years has elapsed since the onset of the initial symptoms without further disturbances. Some children have succumbed to bilateral carotid artery dissections occurring at different times; this may suggest a generalized disorder of the arterial walls such as fibromuscular hyperplasia in some cases.

Angiographic Findings

The wide angiographic spectrum of the disease includes: tapering of the arterial lumen with the typical “string sign” (Cases 1 and 4); “sausage-like” swelling (Case 2) or fusiform dilatation (Case 3) with proximal and distal narrowing of the vessel; a double lumen, and the string-and-pearl sign. The dissection may also progress with further enlargement of the involved vessel or additional aneurysmal dilatation. The finding that spontaneous improvement occurred in fusiform aneurysms both in our and in other reported cases indicates that these cases fare better than those with other angiographic configurations.

Although the differentiation between ruptured arterial dissection and vasospasm is crucial, the vascular changes found in the two conditions may sometimes be similar. Cerebral vasospasm is ordinarily delayed in onset after aneurysmal SAH. We believe that the finding of “arterial narrowing” early after the bleeding suggests a dissection.

In our Case 2 the dilated artery returned angiographically to normal within 3 weeks, and 24 days were
sufficient in Case 4 to document the recanalization of the occluded basilar artery. In cervical carotid artery dissections, recanalization may be present 1 month after the onset of symptoms and this time interval may be sufficient to confirm the evolution of the intracranial segment. Residual fusiform stenosis may depend on the reparative fibrous tissue occupying the false lumen. Other changes including segmental and wedge-shaped narrowing of the healed vessel (Fig. 2 right), noted also in cervical carotid artery dissections, are probably the result of a residual scar. Alternatively, they may represent the original intimal fibro-elastic thickening that some authors consider the major cause of intracranial dissections.

Treatment

Although heparinization is recommended in the management of intracranial arterial dissection, both of our cases of nonhemorrhagic dissection resolved without anticoagulation. A possible drawback of this therapy includes clinical deterioration promoted by mural hemorrhage at the dissection site. Microvascular bypass procedures have been advocated in cases of partial or complete occlusion of the cerebral arteries. The only effective and safe management of ruptured dissecting aneurysms, at least in the vertebral artery, seems to be sacrifice of the parent vessel. However, the results of this treatment are not always satisfactory and fatal SAH has occurred in some surgical patients. Ischemic complications may arise as a consequence of embolization from the clip site or the aneurysm itself or due to the development of an intraluminal thrombus. Other procedures, such as reinforcement of the arterial wall, seem inconclusive and the surgical approach per se may be disastrous. We believe that in a subset of neurologically stable patients the possibility of a spontaneous repair should be investigated before operation, particularly if only palliative surgical treatment is planned.

References


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