Dissection of the Posterior Cerebral Arteries

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Most reported arterial dissections involve the carotid and vertebral arteries in the neck. Within the intracranial arteries, dissections most often involve the vertebral and basilar arteries. Intracranial carotid and middle cerebral artery dissections occur much more rarely and seem to develop in younger individuals compared with patients who develop posterior-circulation intracranial dissections. Dissections that involve the posterior cerebral arteries (PCAs) are extremely uncommon and have been very rarely reported. We now describe 3 patients who developed PCA lesions that were most likely dissections. Two patients had acute strokes caused by PCA dissections, and 1 patient had a chronic dissecting aneurysm of the PCA.

REPORT OF CASES

CASE 1

A 51-year-old psychiatrist became bored and restless during a meeting away from home. He rented a sailboat despite strong storm warnings. The winds became very strong and he felt “battered by the sea.” The sailboat swept out of control and he had to grip strongly to hold on to prevent falling. Later that day, his neck and back of the head hurt. He had a brief vision of a scintillating waterfall to his left. A week later, during which he continued to have an occipital ache, he again had a similar visual illusion to his left. Two days later he developed ptosis and left limb incoordination. Nine years previously, during extreme stress, he had developed a left hemiparesis due to a right putaminal hemorrhage and had first been found to be hypertensive. Blood pressure had been well controlled since.

On examination, his blood pressure was 125/80 mm Hg. Results of cognitive tests, including drawing, copying, and visualization, were normal. Visual fields were normal. The right eyelid was severely ptotic. The eyes diverged, with the right resting down and out. The patient had a left gaze palsy accompanied by tortional nystagmus in a clockwise rotation that was more obvious in the left eye. On attempted down-gaze, the left eye dipped below the right and showed prominent tortional nystagmus. Pupils were 4.5 mm and reacted normally. The left limbs were clumsy but not weak. Left limb reflexes were exaggerated, and the left plantar response was extensor. Gait was wide-based.

A diffusion-weighted magnetic resonance (MR) image (Figure 1A) showed a right paramedian midbrain infarct. A fluid-attenuated inversion recovery MR image (Figure 1B) showed a right paramedian thalamic hyperintensity and a small old right putaminal cavity. The cerebral hemispheres, including the right temporal and occipital lobes, appeared normal. Magnetic resonance angiography (MRA) (Figure 1C) showed a dilated proximal segment of the right PCA followed by a string-like lumen.

The patient’s gait and left-limb clumsiness soon returned to normal, but diplopia and some oscillospia persisted. Re-
A 31-year-old woman had an anxiety attack with chest tightness and rapid heart beating. Several days later, after taking an antidepressant prescribed for her, she had protracted severe vomiting. After vomiting, she suddenly noted a strange feeling in the back of the head on the right, “like a pop or snap.” Immediately thereafter she noted that the left side of her face, left ear, left side of her trunk, and left limbs became numb. She curled up in a dystonic posture. The pop in the head was very transient, and the numbness went away in an hour. During the next hours and days she bumped into objects on her left but was not aware of a visual defect. Three days later, the left numbness returned, lasted 6 hours, and then cleared, leaving only a slight “dullness” in her left hand. She became aware of some difficulty seeing objects to her left, an abnormality that persisted.

At age 23 years, the patient had had cancer of the uterine cervix. She later had 1 pregnancy and had a 2-year-old child at the time of the stroke. She had a history of migraine headaches but did not have a migraine during the onset of her neurologic symptoms.

Results of general examination and vital signs were normal. The patient was able to write, draw, and copy normally and did very well in describing pictures shown to her quickly. She had no left neglect. She had an incongruous hemianopia with a severe, virtually complete, hemianopia in the right eye but only a hemichromatopsia in the left eye. Motor, sensory, and reflex functions were normal, and she walked normally. Magnetic resonance images (Figure 2A and B) showed an infarct that involved the right occipital and temporal lobes and the right ventrolateral thalamus in the distribution of thalamogeniculate branches of the PCA. An MRA (Figure 2C and D) showed a very abnormal right PCA with portions of dilation and narrowing, but the remainder of the extracranial and intracranial arteries were normal. Evaluation for coagulopathy and a cardiac source of embolism was negative. The visual field abnormality persisted, but the patient adapted well. A follow-up MRA (Figure 2E) showed improvement in the stenotic PCA lesion.

A 63-year-old gynecologist developed a headache accompanied by a “feeling of anxiety” and some confusion. He asked his physician to order a computed tomographic scan because he was worried that he had developed cerebral metastases from his known prostatic cancer. An MR image was obtained because the computed tomographic scan was interpreted as abnormal. The patient received no specific treatment. Months later he had several episodes of brief sudden flashes of light that he localized to his right eye. He recognized no persistent visual or other neurologic deficit. When he was younger, he had had severe headaches and photophobia, diagnosed as migraine. These headaches stopped at age 40 years. His family had severe cardiovascular disease.

On examination, his blood pressure was 140/110 mm Hg. Results of general and neurologic examinations were normal except that the patient could not readily name drugs that he often prescribed to his patients and he had a slight right hemihypesthesia. An MR image showed small infarcts in the left thalamus and left temporal lobe. An MRA and contrast angiography (Figure 3) showed a very abnormal left PCA characterized by an aneurysmally dilated artery beginning in the ambient segment. His right sensory symptoms and anomia improved, and he had no subsequent neurologic events. No follow-up brain or vascular imaging was performed.
ment (sailing in a battering wind, and violent pro-
tracted vomiting), the onset with head and neck pain,
the appearance of the PCAs on vascular imaging, and im-
provement with time make it clear that the vascular pro-
cess in these patients was dissection. Patient 2 had no
vascular risk factors for other causes of occlusive dis-
ease. In patient 3, the large aneurysm could conceivably
have been a fusiform aneurysm, but the appearance is
more typical of a dissecting aneurysm. Without surgery
or necropsy, it is impossible to be absolutely certain that
the lesions represented dissections.

An almost constant feature of arterial dissection is pain.
The large arteries that course within the subarachnoid
space are invested with pain-sensitive nerve endings. In
each patient, the tear in the PCA was heralded at the on-
set by an unusual sensation in the head, referred to as a
pop or snap by one of the patients. This was closely fol-
lowed by discomfort and aching that was localized to the
occiput, and occiput and neck, on the side of the dissec-
tion.

In 2 of the patients, the dissection was almost surely
trigged by sudden motion (buffeted by the sea) or ce-
phalic pressure changes (repeated vomiting). Although
extracranial carotid and vertebral artery dissections are
well known to be related to sudden stretching and mo-
tions, precipitation of intracranial dissections by physi-
ical perturbations has not been widely discussed or re-
ported. One of us (L.R.C.) has seen 2 other patients with
intracranial vertebral artery dissections related to physi-
ical perturbation. One young man developed headache
diplopia after stretching for a ball during a volley-
ball game. The second patient was a fireman who was
inadvertently hit forcibly on the side of the head by a large
hose while helping to put out a fire. He had a dissection
that extended from the intracranial vertebral artery into
the basilar artery and became quadriparetic.

All 3 patients in this report had transient symptoms:
hemisensory in 1 patient and visual experiences in 2 pa-
tients. These involved the thalamus and the visual field
supplied by the PCA dissection. Hemianopic visual and
hemisensory symptoms are the most common symp-
toms described in patients who have transient ischemic
attacks related to PCA stenosis.12 One of the patients de-
veloped neurologic signs caused by a midbrain infarct.
An incomplete third nerve palsy, sparing the pupil, and
an ocular tilt reaction were accompanied by clumsiness
of the contralateral limbs probably explained by involvement of extrapyramidal fibers in the medial portion of the cerebral peduncle. The paramedian midbrain and thalamus are supplied by penetrating branches of the proximal portion of the PCA at or very near the origin of the artery from the basilar artery. This is the portion of the artery that was dilated. The transient hemianopic visual symptoms can be explained by hypoperfusion or small emboli entering cortical branches of the PCA.

The second patient had transient hemisensory symptoms. When encountered in patients with PCA disease, the causative lesion is always proximal and before the thalamogeniculate and posterior choroidal artery branches that supply the thalamus. The infarct that developed in this patient was most likely in territory supplied by the lateral posterior choroidal artery as well as in the occipital and temporal lobe territory supplied by the PCA. Visual field defects and hemisensory loss are the most common signs in patients with PCA territory infarction.

All 3 patients had pathologically dilated segments of the PCA. In the third patient the enlargement would qualify as aneurysmal. It is possible that this patient had a congenitally dilated artery that became dissected, or that repeated dissections occurred. In the other 2 patients, regions of both dilatation and narrowing were present. The rapid onset and improvement in the artery on subsequent vascular imaging make it clear that the lesions were dissections.

We were able to find descriptions of 17 patients with PCA dissections. Seven of these patients presented with subarachnoid hemorrhage. For unclear reasons, subarachnoid hemorrhage is an unusual presentation in patients with anterior-circulation intracranial dissections but is common in patients with dissections involving the vertebral and basilar arteries. The patients with subarachnoid hemorrhage included 5 women and 2 men, with an average age of 31 years (range, 13-44 years). The clinical syndrome in these 7 patients, when described, included headache and vomiting but no focal signs at onset. All had aneurysmal dilatation of 1 PCA, usually including the proximal segments of the artery. One patient had a dissecting aneurysm of the temporal branch of the left PCA. Follow-up imaging of 1 patient showed diffuse narrowing of the PCA and an occipital lobe infarct. One patient, a 40-year-old man, had a large PCA aneurysm with an intimal flap detected during an evaluation of headache that followed a minor car accident. This patient was the only one with subarachnoid hemorrhage in whom trauma or physical perturbation was mentioned as a potential precipitant. One patient, a 41-year-old woman, had mixed connective-tissue disease as a possible factor in promoting formation of her PCA aneurysm.

Six previously described patients presented with brain ischemia, including 2 men and 4 women with an average age of 30 years (range, 18-60 years) (Table). Narrowing of the PCA with a string sign and/or occlusion were common findings, but some patients had aneurysmal regions of dilatation. Only one report mentioned a possible physical precipitant: vigorous basketball. Headache was a prominent and early feature. In the 5 patients in whom symptoms and signs were described, all had major clinical signs including hemianopia and hemisensory loss, often accompanied by motor deficits. In the 4 patients in whom brain imaging results were reported, there were large infarcts in the hemispheric and, often, thalamic territory supplied by the PCA.

Intracranial dissections are much less common than extracranial dissections. In addition, many intracranial dissections that cause occlusion of intracranial arteries are not recognized as dissections, especially if they occur in children and young adults. The intracranial

Figure 3. Magnetic resonance imaging studies in patient 3. A, Magnetic resonance angiogram, view from below, showing the proximally aneurysmally dilated posterior cerebral artery (PCA) (paired arrows) with distal narrowing (single arrow). B, Cerebral angiogram, anteroposterior view, showing a very large dilated left PCA (arrow).
Anterior circulation is the site of dissection more often in young adults, while posterior-circulation intracranial arterial dissection more often occurs in older patients. A hemodynamic mechanism with hypoperfusion is more likely to cause brain ischemia in patients with intracranial dissections, while embolism of thrombus is the most common cause of ischemia in patients with extracranial carotid and vertebral artery dissections. The role of congenital and inherited abnormalities of connective tissue and collagen in contributing to intracranial arterial dissections is now unknown.

Treatment of intracranial dissections is controversial. Some intracranial dissections cause brain ischemia by hypoperfusion distal to an intramural hematoma that compromises the arterial lumen. Augmenting blood volume and blood pressure might increase perfusion to the ischemic area through collateral vessels. Some intracranial dissections cause infarction in relation to thrombus formation, propagation, and embolization within the lumen of the dissected artery. This is especially important in patients with dissecting aneurysms (as in patient 3), since inefficient antegrade flow encourages stasis and thrombus formation. Anticoagulation with heparin and, later, warfarin could theoretically prevent thrombus formation and embolization. However, intracranial dissections and fusiform aneurysms can rupture, causing severe parenchymatous and/or subarachnoid bleeding, especially if the dissection extends into the arterial adventitia. Agents that compromise platelet function would also enhance the severity of bleeding, should rupture occur, and have less theoretical benefit against red clot formation. Treating physicians must weigh the benefit-risk ratio of anticoagulants in each individual patient with intracranial dissection. Stenting with or without preceding angioplasty is another potential treatment for intracranial dissections that cause stenosis of arteries, although there is little present experience with this technique in this condition, and recovery of luminal diameter is the rule without mechanical intervention.

Dissections can involve the main intracranial arteries and their branches. Physical perturbations may play an etiologic role just as they do in dissections of neck arteries. Headache is a prominent and early symptom. As in our third patient, neurologic symptoms can be delayed days or weeks after development of the arterial dissection. Dissections either involve the medial-adventitial region of the arteries, causing aneurysmal dilation and often subarachnoid hemorrhage, or affect the intima-media layers, causing arterial narrowing and thrombus formation with resultant ischemia in regions supplied by the dissected artery.

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Table. Patients With PCA Dissections and Brain Ischemia

<table>
<thead>
<tr>
<th>Source</th>
<th>Age, y/ Sex</th>
<th>Angioimaging</th>
<th>Symptoms/Signs</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Present report</td>
<td>51/M</td>
<td>Dilated proximal R PCA, then irregular narrowing; midbrain and thalamic infarct</td>
<td>Headache; then transient visual, then R III nerve palsy, ocular tilt, L hemiataxia</td>
<td>Antiplatelet agent</td>
<td>Diplopia, nystagmus</td>
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<tr>
<td></td>
<td>31/F</td>
<td>Dilated proximal R PCA, then narrowing; R thalamic and tempo-occipital infarct</td>
<td>Headache; then transient L hemisensory, L hemianopia</td>
<td>Heparin, warfarin sodium</td>
<td>L hemianopia</td>
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<tr>
<td></td>
<td>63/M</td>
<td>Aneurysm in distal L PCA</td>
<td>Headache; transient visual experience</td>
<td>Antiplatelet agent</td>
<td>Asymptomatic</td>
</tr>
<tr>
<td>Le Tu et al³</td>
<td>26/F</td>
<td>Proximal L PCA narrowed, then dilated, then occluded; L thalamic and occipital infarct</td>
<td>Headache; then R hemianopia, R hemisensory, R clumsiness</td>
<td>Heparin, warfarin</td>
<td>R upper quadrantanopia</td>
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<tr>
<td></td>
<td>20/F</td>
<td>R PCA narrowed, then occluded; R thalamic and occipital infarct</td>
<td>Headache; then L hemianopia, L hemisensory, left clumsiness</td>
<td>Heparin, warfarin</td>
<td>NR</td>
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<tr>
<td>Fisher et al²</td>
<td>18/M</td>
<td>R PCA narrowed</td>
<td>Neck pain; then L hemianopia, L hemisensory loss</td>
<td>NR</td>
<td>L hemianopia</td>
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<tr>
<td>Pozzati et al⁷</td>
<td>60/M</td>
<td>PCA string sign</td>
<td>NR</td>
<td>NR</td>
<td>“Recurrent symptoms”</td>
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<tr>
<td>Berthier and Bourrat¹⁰</td>
<td>24/F</td>
<td>L proximal PCA fusiform, dilated, double lumen</td>
<td>Headache; then R hemianopia, hemisensory, hemiataxia, amnesia</td>
<td>Heparin, warfarin</td>
<td>Hemianopia, amnesia</td>
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<tr>
<td>Maillo et al¹¹</td>
<td>33/F</td>
<td>R PCA aneurysmal dilation, occluded</td>
<td>Headache; then L hemianopia, L hemisensory, L hemiparesis</td>
<td>NR</td>
<td>L hemianopia</td>
</tr>
</tbody>
</table>

Abbreviations: L, left; NR, not reported; PCA, posterior cerebral artery; R, right.


14. Le Tu PT, Zuber M, Meder JF, Maciel et al. Founder Haplotype for Machado-Joseph Disease in the Indian Population,” published in the April 2005 issue of the ARCHIVES (2005;62:637-640), in the Table at the bottom of page 639, the haplotype percentages in both Indian columns were incorrect. The table is reprinted here with the correct values.

**Table. Haplotypes in Normal and Expanded Alleles in the Indian and Azorean Populations***

<table>
<thead>
<tr>
<th>No. of CAG Repeats</th>
<th>ACA</th>
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<th>GGC</th>
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<tbody>
<tr>
<td></td>
<td>Indian</td>
<td>Azorean</td>
<td>Indian</td>
<td>Azorean</td>
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<tr>
<td>14-21</td>
<td>10.3</td>
<td>5</td>
<td>56.4</td>
<td>68</td>
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<tr>
<td>22-26</td>
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<td>27-37</td>
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<td>7</td>
<td>9.6</td>
<td>71</td>
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<tr>
<td>Expanded</td>
<td>100</td>
<td>47</td>
<td>0</td>
<td>45</td>
</tr>
</tbody>
</table>

*Data for the Azorean population are from Maciel et al.8
†In normal alleles in the Indian population there were 161 chromosomes.

In addition, a word was misplaced in the acknowledgments on page 640 near the top of column 2. The acknowledgments should have read as follows: “Acknowledgment: We thank Samir K. Brahmachari, PhD, for providing intellectual support during the course of this investigation; Vani Brahmachari, PhD, for critical evaluation of the manuscript; and Sangeta Sharma, BSc, for help with the GeneScan and sequence analysis.”