Perimesencephalic Subarachnoid Hemorrhage
Additional Perspectives From Four Cases

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Background Nonaneurysmal perimesencephalic hemorrhage, a distinct form of subarachnoid hemorrhage, is a recently described variant of intracranial hemorrhage. We describe two patients who presented with unusual features of this type of subarachnoid hemorrhage and also two patients who had a perimesencephalic pattern of hemorrhage due to a ruptured posterior circulation aneurysm.

Case Descriptions The first patient, a 41-year-old woman with perimesencephalic hemorrhage, underwent an exploratory craniotomy because angiography had suggested an anomaly of the basilar tip. No source of hemorrhage could be identified at the time of surgery. The second patient was a 3-year-old boy who presented with opisthotonos and who was found to have a perimesencephalic hemorrhage. Angiography revealed no source for the hemorrhage. The third patient, a 54-year-old man, had a perimesencephalic pattern of subarachnoid hemorrhage from a vertebrobasilar junction aneurysm.

In approximately 15% of patients with spontaneous subarachnoid hemorrhage (SAH) the cause of the hemorrhage cannot be detected despite detailed imaging studies. It has repeatedly been shown that SAH of unknown origin is associated with a much better outcome than aneurysmal SAH, although recurrent bleeding and delayed cerebral ischemia do occur. Recently, van Gijn and colleagues described a benign variant of SAH, the so-called perimesencephalic nonaneurysmal hemorrhage. This type of SAH is characterized radiographically by a pattern of hemorrhage restricted to the perimesencephalic or prepontine cisterns in combination with a normal angiogram and clinically by an invariably excellent prognosis.

Since its original description, it has subsequently been shown that the good prognosis of SAH of unknown etiology can be largely explained by the high prevalence of perimesencephalic nonaneurysmal hemorrhage in series of patients with SAH and a negative angiogram. Thus, when perimesencephalic-type hemorrhages are excluded from series of patients with SAH and a negative angiogram, outcome is much less favorable. Despite the increasing number of reports of series of patients with nonaneurysmal perimesencephalic hemorrhage, many uncertainties remain regarding this distinct type of SAH, especially relating to their etiology and evaluation. In this communication we describe four patients who manifest some unusual aspects of perimesencephalic SAH. Two patients had a nonaneurysmal perimesencephalic hemorrhage, and two had a perimesencephalic hemorrhage due to a ruptured posterior circulation aneurysm.

Case Reports

Patient 1

A 41-year-old woman developed the acute onset of a severe headache associated with nausea and vomiting. A computed tomography (CT) scan, obtained 2 hours after the ictus, showed an SAH limited to the interpeduncular and prepontine cisterns with minimal extension into both sylvian fissures. She was transferred to our institution.

On admission, examination only showed moderate nuchal rigidity. Four-vessel cerebral angiography showed a small domelike dilatation of the basilar tip but was otherwise normal (Fig 1). Magnetic resonance imaging (MRI) only showed residual blood in the interpeduncular cistern. Angiography was repeated 7 days later and was unchanged. Subsequently, through a right frontotemporal craniotomy, the upper basilar artery was explored, and a small bulbous dilatation was identified posteriorly at the basilar tip. There was no evidence that this dilatation had ever ruptured or leaked, and no other suspicious areas could be identified. The patient recovered well from her surgery and has remained asymptomatic during 1 year of follow-up.
Patient 1

A 3-year-old white boy developed the acute onset of a severe headache accompanied by nausea and vomiting. Twenty-four hours later marked neck stiffness was noted by his parents. Lumbar puncture revealed grossly hemorrhagic cerebrospinal fluid, and examination of the supernatant revealed xanthochromia. He was transferred to our institution.

The patient was a product of an uncomplicated full-term pregnancy. There was no history of trauma or evidence of child abuse.

On admission, examination was remarkable only for marked nuchal rigidity and opisthotonos. Laboratory examinations, including complete blood count, serum electrolytes, liver function tests, coagulation studies, and drug screen, were all within normal limits. Review of the outside CT scan revealed a localized hemorrhage within the suprasellar and prepontine cisterns (Fig 2). MRI revealed the area of acute hemorrhage within the prepontine cistern extending rostrally into the interpeduncular and suprasellar cisterns, separate from the basilar artery (Fig 3). Four-vessel cerebral angiography only revealed narrowing of the basilar artery, consistent with vasospasm secondary to the SAH. An MRI examination of the entire spine was normal. A second four-vessel cerebral angiogram 10 days later was normal, with resolution of the vasospasm. Neuropsychological evaluation was normal. The patient was discharged home in excellent condition 2 weeks after admission.

The patient has continued to do well during 10 months of follow-up. A second MRI of the head with gadolinium administration was unremarkable (Fig 3).

Patient 3

This 54-year-old man developed a sudden severe headache associated with nausea and vomiting. CT
FIG 4. Patient 3. Axial computed tomographic scan, obtained 6 hours after the ictus, reveals a subarachnoid hemorrhage limited to the interpeduncular and right ambient cisterns. The leaves of the tentorium may be more dense than normal, indicating the presence of a thin layer of subarachnoid blood.

examination showed a localized hemorrhage in the preopticine and interpeduncular cisterns with no extension into the frontal interhemispheric or sylvian fissures (Fig 4). No intraventricular or parenchymal hemorrhage was noted. The patient was transferred to our institution.

Examination was remarkable only for moderate nuchal rigidity. A four-vessel cerebral angiogram was interpreted as normal (Fig 5A). MRI examination of the cervical spine was normal. One week later, a second angiography revealed a 3-mm aneurysm rising between two limbs of a fenestration of the proximal basilar trunk (Fig 5B). In retrospect, the aneurysm was present on the initial study. The aneurysm was successfully treated with a single Guglielmi detachable coil measuring 3 mm x 4 cm.

Patient 4

This 43-year-old man suddenly developed a severe headache after an orgasm. CT examination, performed 3 hours after the onset of symptoms, showed an SAH centered within the right ambient cistern with no extension into the frontal interhemispheric or sylvian fissures (Fig 6). No intraventricular or parenchymal hemorrhage was seen. Two cerebral angiograms were reported as normal. The patient was transferred to our institution.

Examination showed mild nuchal rigidity only. On review of the angiograms, a 2-mm right posterior cerebral artery aneurysm (P2 segment) was noted (Fig 7). Through a right frontotemporal craniotomy a very
thin-walled aneurysm was encountered, which rose at the P2 segment along with the origin of a circummesencephalic branch. There was clear evidence of recent rupture, and the aneurysm was clipped. The patient recovered well from his surgery and has not had recurrent problems.

Discussion

Nonaneurysmal perimesencephalic hemorrhage has become well recognized as a distinct type of SAH and may account for up to two thirds of all SAHs of unknown cause. Patients with a typical clinical picture of perimesencephalic SAH are adults with the acute onset of headache without loss of consciousness or focal neurological symptoms. Neurological examination is normal, and often only meningeal irritation is found. The clinical course is without secondary deterioration from delayed cerebral ischemia or recurrent bleeding, and the long-term prognosis is invariably excellent. Perimesencephalic SAH is diagnosed on the basis of CT or MRI examination, which demonstrates a localized area of hemorrhage centered within the perimesencephalic or prepontine cisterns without intracerebral or intraventricular extension. After the term “perimesencephalic hemorrhage” was coined in 1985, it has become apparent that a number of these hemorrhages are centered around the prepontine cistern and that some also extend into the suprasellar cistern. With the advent of MRI, blood may also be demonstrated anterior to the medulla oblongata. Angiography in perimesencephalic hemorrhage rarely reveals an intracranial aneurysm or other source of bleeding. However, full cerebral angiography remains mandatory in all patients with perimesencephalic SAH because a ruptured basilar artery aneurysm may produce a similar pattern of hemorrhage on imaging studies. Repeated angiography after a negative study has not demonstrated an aneurysm or other source of hemorrhage in previously reported series but sporadically has resulted in cerebral infarction. Therefore, it has been suggested that it is reasonable to limit angiography to a single examination. However, in 2% to 16% of patients with a pattern of perimesencephalic hemorrhage, a posterior circulation aneurysm is found. Furthermore, it is well known that angiography may not demonstrate a ruptured aneurysm in all cases of aneurysmal SAH on the initial examination. The posterior circulation aneurysms in our patients 3 and 4 were difficult to recognize on the initial angiograms. This failure to identify a posterior circulation aneurysm highlights the concerns in clinical practice when a patient with a perimesencephalic pattern of SAH is encountered. Possibly, physicians who recognize a perimesencephalic pattern of SAH on CT may bias their expectations for a negative angiogram. Although our case material does not support a second angiographic study in patients with perimesencephalic hemorrhage, it does underscore the need for a high index of suspicion for a ruptured aneurysm of the posterior circulation. Moreover, a perimesencephalic pattern of SAH may be caused not only by a ruptured aneurysm of the basilar top but also by those in more unusual...
locations, such as the distal posterior cerebral artery or verteobasilar artery junction.

Nonaneurysmal perimesencephalic hemorrhage has not been described in childhood or adolescence. The youngest reported patient with nonaneurysmal perimesencephalic hemorrhage was 22 years. The clinical and radiographic characteristics of our 3-year-old patient with SAH were consistent with nonaneurysmal perimesencephalic hemorrhage. Other causes of SAH in this age group, eg, arteriovenous malformation, aneurysm, blood dyscrasia, brain tumor, intracranial arterial dissection, sickle cell disease, or structural spinal disorders, were excluded in our patient. Battery could possibly have created a similar pattern of hemorrhage, but there was no evidence of child abuse in our patient. Moreover, isolated SAH in the absence of retinal hemorrhages or subdural hematoma is rare in the battered child syndrome. Furthermore, if SAH due to battery is identified it is often in the interhemispheric fissure. Recognition of nonaneurysmal perimesencephalic hemorrhage is important in the pediatric as well as the adult population, especially in view of its benign nature. Restrictions of activities or other adjustments in lifestyle do not need to be instituted in patients with nonaneurysmal perimesencephalic SAH, certainly a pertinent point for an active young child. Whether the long-term clinical course in children is as favorable as that in adults, however, remains to be determined, but there are no compelling arguments to believe otherwise.

The clinical entity of nonaneurysmal perimesencephalic hemorrhage has not been defined by a pathologic substrate, but a venous or capillary source of bleeding has been implicated. An exploratory craniotomy in one of our patients failed to disclose a source of hemorrhage despite the presence of an irregular basilar tip. Likewise, others have described patients with nonaneurysmal perimesencephalic hemorrhage who underwent an exploratory craniotomy without elucidating the cause of the hemorrhage. These negative exploratory craniotomies suggest that the source of the hemorrhage is probably not arterial.

The patients described in this report demonstrate several unusual aspects of perimesencephalic hemorrhage and suggest that the clinical spectrum of this type of SAH needs further refinement. The index of suspicion for a posterior circulation aneurysm should remain high in patients who present with a perimesencephalic pattern of SAH. The diagnosis of nonaneurysmal perimesencephalic hemorrhage is one of exclusion.

References

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*Stroke*. 1994;25:1507-1511
doi: 10.1161/01.STR.25.7.1507

*Stroke* is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0039-2499. Online ISSN: 1524-4628

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World Wide Web at:
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