**Case Reports**

**Subarachnoid Hemorrhage of Unknown Etiology along the Cortical Convexity**

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**Abstract**

**Background:** Only 8% to 22% of cases of subarachnoid hemorrhage (SAH) are of nonaneurysmal origin. Among these, perimesencephalic nonaneurysmal SAH is a distinct clinical and radiologic entity with normal angiographic findings and a good prognosis. In contrast, SAH of nonaneurysmal origin occurring along the cortical convexity is rare and poorly understood. We report 2 cases of subarachnoid hemorrhage along the cortical convexity and discuss their possible etiologies.

**Methods:** In a retrospective analysis of 234 patients with SAH, we identified 2 patients with a typical computed tomographic pattern of convexity SAH that was associated with no known etiology.

**Results:** In these 2 cases, the source of hemorrhage could not be identified with computed tomography, magnetic resonance imaging, or digital subtraction angiography, although neurovascular outcomes were good. The patients reported such incidents as coughing or exertion immediately before headache developed. These incidents may have caused increased intracranial pressure.

**Conclusion:** We suggest the possible involvement of a brief increase in intracranial pressure, such as that accompanying coughing or exertion, in the occurrence of SAH along the cortical convexity.


**Key words:** subarachnoid hemorrhage, cortical convexity, etiology

**Introduction**

In 8% to 22% of patients with subarachnoid hemorrhage (SAH), no known etiology, such as aneurysm or vascular malformation, is evident from initial digital subtraction angiography (DSA). In addition to DSA, magnetic resonance imaging (MRI) of the brain and spine is often performed to exclude small vascular abnormalities, such as arteriovenous malformation, cavernous angioma, and venous anomalies, particularly those at the craniocervical junction. A previous study has found that 3-dimensional (3D) computed tomographic (CT) angiography is useful in cases in which initial angiograms fails to demonstrate the source of SAH.
However, in approximately 5% to 34% of patients with nontraumatic SAH, no obvious source of hemorrhage can be demonstrated with a combination of DSA, MRI, and 3-D CT angiography. Among SAHs of unknown etiology, one subset designated as perimesencephalic SAH has been diagnosed with the findings of initial CT findings and the absence of abnormalities on vascular imaging. In a smaller number of patients with SAH of unknown etiology according to initial DSA findings, initial CT revealed a circumscribed SAH along the cortical convexity. In most of these cases, the cause of SAH—such as postpartum eclampsia, brain tumor, coagulopathy, reversible encephalopathy, carotid stenosis, and cerebral venous sinus thrombosis—was ultimately identified. Recently, Refai et al. reported 20 cases of spontaneous isolated convexity SAH. The cause of hemorrhage was identified in 13 of these cases but remained unknown in 7 cases. Another 5 patients with SAH along the cortical convexity, which was diagnosed with initial CT but whose origin could not be determined with DSA or MRI, have been described by Spitzer et al. and Patel et al. In the present paper, we describe 2 patients with this type of SAH and discuss their clinical course and the most likely causes of their condition.

Methods

Of the 234 patients treated for spontaneous SAH at our institution from 1999 through 2010, 12 (6.1%) had an SAH that had been diagnosed with initial CT but whose etiology could not be determined with MRI and DSA (including that of the internal and external carotid artery and the bilateral vertebral arteries); 2 (0.9%) of these SAHs of unknown origin were along the cortical convexity. Exclusion criteria included coagulopathy, including that due to antiplatelet or anticoagulant drugs, hematological or liver function disorders, hypertensive encephalopathy, and reversible posterior encephalopathy syndrome. The clinical characteristics of the present 2 cases (Table) and the 12 previously reported cases are discussed below.

Case Illustrations

Case 1

A 54-year-old woman with a history of hypertension presented with a severe headache that had developed suddenly after a coughing episode. On examination, she was conscious and fully oriented to time, place, and person. General and neurological examinations identified no abnormalities, except for the complaint of headache. Emergency CT disclosed hyperdensity in the subarachnoid space, sparing the basal cisterns and involving the sulci of the left cerebral convexity (Fig. 1A). Results of cerebrospinal fluid analysis were unremarkable, except for a finding of 98 red blood cells (RBCs)/mm³. DSA revealed no abnormalities, such as sinus thrombosis and abnormal vascular stenosis in the arterial or venous phase, and thus found no potential cause of SAH (Fig. 1B). Results of MRI, including gradient-echo T2-weighted images, fluid-attenuated inversion recovery sequences, diffusion-weighted sequences, and 3D time of flight MR angiography of the intracranial arteries, were all normal without SAH. The patient was treated conservatively and showed complete radiologic and clinical recovery at follow-up. Coagulation studies, including tests for antithrombin III, protein S, protein C, anti-

<table>
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F: female; GR: good recovery; HA: headache; HT: hypertension; M: male; VS: vegetative state

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Table Summary of cortical subarachnoid hemorrhage of unknown etiology in this report

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phospholipid antibodies, and lytic state, were negative.

Case 2
A 68-year-old man with a history of hyperlipidemia presented with a severe headache that had suddenly developed while he was vigorously sawing a tree. On examination, he was conscious and fully oriented to time, place, and person. General and neurological examinations identified no abnormalities other than a headache. Findings on emergency plain CT demonstrated hyperdensity (Fig. 2A), sparing the basal cisterns and involving the sulci of the right cerebral convexity. Results of a cerebrospinal fluid study were unremarkable, except for the finding of 693 RBCs/mm³. DSA revealed no abnormalities (Fig. 2B) in the arterial and venous phases and did not reveal any potential cause of the SAH. Findings were normal on additional MR imaging, including gradient-echo T2-weighted images, fluid-attenuated inversion recovery sequences, diffusion-weighted sequences, and 3D time of flight MR angiography of the intracranial arteries. The patient was treated
conservatively and showed complete radiologic and clinical recovery at follow-up. The results of blood tests, including those for protein C, anti-cardiolipin antibody, and lytic state, were negative.

Results

Six of 14 reported cases of SAH of unknown etiology (including those of our 2 cases) occurred in men. The patients ranged in age from 35 to 91 years, with a mean age of 69.8 years. All but 1 cortical SAH were located on the unilateral frontoparietal convexity around the central sulcus. With regard to the initial symptoms, 6 patients had headache only, while the remaining 8 patients had other neurologic symptoms, such as monoplegia, numbness, and seizures. In many cases, neurological symptoms, such as monoplegia and numbness, were transient and short-lived (10 to 60 minutes). All 14 cases had favorable outcomes without hydrocephalus. In the reports of 12 of the cases, no details were provided about the circumstances at onset or the possible trigger.

Discussion

In these cases of SAH of unknown etiology along the cortical convexity, the source of hemorrhage was not recognized with CT, MRI, or DSA of arterial and venous structures. In addition, both of our patients had reported an incident, such as coughing or exertion, that might have increased intracranial pressure immediately before headache developed. This briefly increased intracranial pressure could have been the cause of SAH.

Etiology of SAH along the Cortical Convexity

Several authors have discussed the etiology of SAH along the cortical convexity. Oppenheinm et al.\(^9\) reported 4 cases of SAH that involved the sulci of the convexity and spared the basal cisterns, as was seen in both of our cases. The etiology of SAH in all cases reported by Oppenheinm et al.\(^9\) was dural sinus thrombosis. Spitzer et al.\(^10\) have reported 12 cases of SAH along the convexity. In these cases, MRI and DSA revealed the following causes: encephalopathy (n=3), cerebral vasculitis (n=2), dural sinus thrombosis (n=2), cortical venous thrombosis (n=1), intracerebral abscesses (n=1), and cerebral cavernoma (n=1). Only 2 of their cases were of unknown etiology. Refai et al.\(^11\) have also reported 20 cases of convexity SAH; the underlying cause was identified in 13 cases. Patel et al.\(^12\) also reported 5 cases of convexity SAH; the underlying cause of hemorrhage was identified in 2 cases. In our cases, DSA and MRI revealed no abnormalities that could indicate the origin of SAH.

Headache developed in our first patient after an episode of coughing and in our second patient after exertion. Benign exertional headache (BEH) and the headaches in our patients have different characteristics. The headaches in our patients appear to be due to SAH. However, we discussed the possibility of a common cause. Tinel\(^13\) first described a cough headache in 1932. Four cases of intermittent paroxysmal headache after exertion were reported. Benign cough headache\(^27\) was considered a subcategory of BEH, which is specifically brought on by physical exertion\(^25\) such as running, coughing, or heavy lifting. It lasts for 5 minutes to 24 hours and manifests at onset as a bilateral throbbing headache. Several theories regarding the origin of BEH have been proposed\(^26\), but none have been proved. The etiology of BEH may be related to pathophysiologic or structural phenomena or to differences in intracranial and spinal pressures.\(^28\) Williams\(^33\) has reported that BEH results from craniospinal pressure dissociation. Cerebral intraventricular and lumbar intrathecal pressures were monitored during and after coughing in 2 patients with BEH. These variables were also measured after the patients performed the Valsalva maneuver while seated upright. Pressure was higher in the ventricle than in the lumbar space after both coughing and the Valsalva maneuver, leading Williams to propose that a pressure differential associated with craniospinal pressure dissociation was the most likely etiology of BEH.

Several articles have proposed a vascular origin for BEH\(^25\). Because exertion increases systemic blood pressure or venous pressure or both, intracranial pressure may also increase with
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exertion. Silbert et al.\textsuperscript{33} have reported cerebral angiography findings of benign sexual headache and BEH occurring sequentially in the same patient. Angiography also demonstrated multiple vasospasms. In our patients, however, angiography findings were normal. We hypothesized that exertion related to increased venous or arterial pressure or acceleration related to coughing or both may have led to breakthrough hemorrhage from fragile veins in the cerebral cortex. Refai et al.\textsuperscript{1}, Patel et al.\textsuperscript{7}, and Spitzer et al.\textsuperscript{8} have also reported cortical SAH of unknown etiology, but they did not report the circumstances at onset. Further study of the details of onset for additional cases of SAH of unknown etiology along the cortical convexity is important to clarify the role of exertion in hemorrhage onset.

References

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