Spontaneous isolated convexity subarachnoid hemorrhage: presentation, radiological findings, differential diagnosis, and clinical course

Clinical article

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Object. The clinical characteristics and overall outcome in patients with spontaneous isolated convexity subarachnoid hemorrhage (SAH) are not well described in the literature. The purpose of this study was to examine the mode of presentation, common origins, radiographic findings, and clinical course in a large case series of such patients.

Methods. A retrospective single-center chart review of all patients in whom nontraumatic primary convexity SAH was diagnosed between 2002 and 2007 was performed. Twenty patients were identified and analyzed for presenting symptoms, radiological and laboratory findings, hospital course, and outcome.

Results. There were 15 women and 5 men in our series, and the mean age was 52 years (range 18–86 years). The most common presenting symptom was headache, with 15 patients experiencing it as a chief complaint. Other frequent manifestations included altered mental status (8 patients), focal neurological deficits (7), and seizure (4 patients). An underlying cause of the hemorrhage was identified in 13 cases, whereas the remainder went unresolved. Of the known causes, 5 were due to posterior reversible encephalopathy syndrome, 3 were caused by thrombocytopenia or anticoagulation, and the remainder were isolated cases of lupus vasculitis, drug-induced vasculopathy, postpartum cerebral angiopathy, hypertensive microangiopathy, and Call–Fleming syndrome. All patients with unknown disease origins had favorable outcomes, whereas 8 of 13 patients with an identifiable underlying disorder experienced favorable outcomes.

Conclusions. Spontaneous isolated convexity SAH is rarely caused by aneurysm rupture, has a distinct mode of presentation, and generally carries a more favorable prognosis than that of aneurysmal SAH. (DOI: 10.3171.JNS.2008.109.12.1034)

Key Words • convexity subarachnoid hemorrhage • nontraumatic subarachnoid hemorrhage • spontaneous subarachnoid hemorrhage

The incidence of spontaneous SAH in the general population is approximately 6–8 cases per 100,000 person-years, of which 85% are the result of a ruptured saccular aneurysm. In 10% of cases, no specific structural lesion or other underlying condition is identified. This group includes the entity known as benign perimesencephalic SAH. The remaining 5% are caused by a variety of conditions, including the following: transmural arterial dissection, cerebral AVM, dural AVF, cavernous malformation, mycotic aneurysm, sinus or cortical venous thrombosis, thrombocytopenia or coagulopathy, vasculitis, cocaine abuse, brain neoplasm, brain abscess, hypertensive microangiopathy, postpartum cerebral angiopathy, and PRES. Although distribution of blood is dependent on the specific site of rupture, the majority of patients with spontaneous SAH demonstrate blood within the basal cisterns, which may extend into the sulci along the cerebral convexities. Isolated SAH along the cerebral convexities, on the other hand, is rare, and has been described in the literature mainly in isolated case reports as well as a recent case series. Although the underlying cause is varied, spontaneous isolated convexity SAHs as a group share many clinical features and neuroradiological characteristics that differ substantially from those of classical aneurysmal SAH. The purpose of the present study is to define these clinical features better, to examine the underlying causes, and to assess outcome in a large series of patients with this uncommon hemorrhagic disorder.

Methods

After obtaining institutional approval from the com-
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mittee on human research, a search of the radiology database at our medical center for “convexity subarachnoid hemorrhage” yielded 864 patients treated between 2002 and 2007. The radiology reports for these patients were then reviewed. We excluded patients with traumatic causes of SAH (459 individuals), patients having intraparenchymal or intraventricular hemorrhagic components (190), and patients having cisternal or sylvian fissure blood (195). The data for the remaining 20 patients were reviewed for this report. These patients with nontraumatic, isolated convexity SAH were analyzed for presenting symptoms, radiological findings, diagnostic studies, causes, treatment, and outcome.

Results

Clinical Presentation

Fifteen of the 20 patients were female; their ages ranged from 18 to 86 years, with a mean age of 52 years. Tables 1 and 2 provide details of the presenting symptoms, radiological findings, diagnostic studies, causes, treatment, and outcomes for these 20 cases. The most common presenting symptom was headache, with 15 patients experiencing headache as a chief complaint. Of these, only 6 experienced a true thunderclap headache and only 1 had neck stiffness. Eight patients had altered mental status, 7 had focal neurological deficits such as unilateral weakness, numbness, or homonymous hemianopia, and 4 patients had seizures (2 focal motor and 2 generalized tonic–clonic).

Radiological Findings

Seven hemorrhages were located over the frontal convexity, 6 over the parietal convexity, and 4 in the frontoparietal region. Two of these 17 patients had bilateral subarachnoid blood. Of the 3 remaining patients, 1 had parietooccipital SAH, 1 had frontotemporal SAH, and 1 had multifocal SAH. Eighteen cases were diagnosed based on CT findings, and 2 were identified via MR imaging. Eleven patients underwent DS angiography as part of their diagnostic workup. No aneurysms, AVMs, dural sinus or cortical venous thromboses were identified by DS angiography, but angiographic evidence of vasculitis or vasculopathy was noted in 3 cases. Seven patients underwent CSF analysis, and all underwent routine blood screening.

Underlying Causes

A cause of the convexity SAH was determined in 13 cases, whereas the remaining seven went unresolved. Five cases were the result of PRES, 3 of which occurred postpartum and 2 of which occurred in the context of immunosuppression. Three cases were caused by thrombocytopenia or coagulopathy: 1 due to thrombocytopenia secondary to end-stage liver failure, 1 due to warfarin-induced coagulopathy related to atrial fibrillation therapy, and 1 due to thrombocytopenia and warfarin-induced coagulopathy related to mechanical heart valve therapy. Other causes included 2 cases of angiographically evident vasculopathy (1 ascribed to postpartum cerebral angiopathy and 1 to drug-induced vasculopathy from the dietary supplement ephedra), 1 case of lupus vasculitis, 1 of hypertensive microangiopathy, and 1 related to Call–Fleming syndrome. Other pertinent findings included anemia in 11 (55%) of 20 patients and a history of hypertension in 7 (35%) of 20 patients.

Clinical Course

General treatment measures taken for most patients included the following: 1) transfer to the neurology/neurosurgery intensive care unit for close observation; 2) anticonvulsant administration for seizure prophylaxis or therapy; and 3) analgesic administration for headache management. A minority of patients (5 of 20) were also given oral nimodipine for vasospasm prophylaxis. Further therapy was tailored to the particular clinical scenario, with treatment specific to the disease’s cause instituted when possible. Patients with PRES were treated with anti hypertensive therapy, anticonvulsants, and steroids if needed. Patients with thrombocytopenia were administered platelets, and patients with coagulopathy were treated via cessation of warfarin therapy and reversal with blood products. Documented hypertension was treated with antihypertensive therapy, lupus vasculitis was treated with steroids, and Call–Fleming syndrome was treated with steroids and analgesic agents.

The hospital course was complicated by SAH-induced hydrocephalus and cerebral vasospasm in only 1 of 20 cases; a 5% incidence rate for either acute hydrocephalus or symptomatic vasospasm. This patient (Case 8) had a large volume of isolated convexity SAH and developed symptomatic hydrocephalus that required ventriculostomy placement, but did not require a permanent shunt. This same patient also developed symptomatic cerebral vasospasm (angiographically proven) in delayed fashion, which was successfully treated with nimodipine and hemodynamic augmentation.

Overall, 15 of 20 patients experienced favorable outcomes, with complete or near-complete resolution of presenting symptoms by the time of discharge (mean hospital stay 16.5 days). The remaining 5 patients had unfavorable outcomes, including 2 with worsened neurological status and 3 deaths. Specifically, 1 patient (Case 15) who had both thrombocytopenia and coagulopathy developed an intraparenchymal hemorrhage leading to a worsened neurological condition. One patient (Case 13) with Call–Fleming syndrome developed intraparenchymal hemorrhage and multiple infarcts leading to a worsened neurological condition. One patient (Case 10) with immunosuppressive PRES developed severe cerebral edema leading to herniation and brain death. The remaining 2 deaths were nonneurological in nature, with 1 patient (Case 14) dying due to complications of leukemia and the other (Case 6) dying of sepsis secondary to a preexisting wound infection. All poor outcomes occurred in patients in whom underlying causative disorders were diagnosed, whereas patients without identifiable underlying causes all had favorable outcomes.

Discussion

Clinical Presentation

Several important observations emerge from this series, which is the largest reported to date. First, patients with spontaneous isolated convexity SAH resembled those with aneurysmal SAH in age and sex distribution, 2 but
their presenting symptoms were atypical. Only 6 patients (30%) in our series presented with thunderclap headache and 1 (5%) had nuchal rigidity. In contrast, patients with aneurysmal SAH present with thunderclap headache in up to 74% of cases and display nuchal rigidity in 35% of cases.8,9 Conversely, seizures and focal neurological deficits were more frequent after spontaneous convexity SAH. Seizure at presentation was noted in 4 (20%) of 20 patients in our series and in 7 (58%) of 12 patients in the series reported by Spitzer et al.40 Both rates contrast with the 6–7% incidence reported after aneurysmal SAH.3,23,31,43

TABLE 1
Overview of imaging and laboratory values in 20 patients with isolated convexity SAH*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Presenting Symptoms</th>
<th>CT Scan</th>
<th>MRI/MRA</th>
<th>DSA</th>
<th>CSF Analysis</th>
<th>Blood Screen</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>18, F</td>
<td>seizures, AMS</td>
<td>rt frontoparietal convexity SAH</td>
<td>rt frontoparietal SAH</td>
<td>T2 (FLAIR)</td>
<td>ND</td>
<td>↑RBCs, ↑WBCs, ↑protein, ↑IgG</td>
</tr>
<tr>
<td>2</td>
<td>74, F</td>
<td>sudden severe HA, nausea, mild nuchal stiffness</td>
<td>rt convexity SAH</td>
<td>rt parietal convexity SAH</td>
<td>T2 (FLAIR)</td>
<td>ND</td>
<td>rt MCA atherosclerosis</td>
</tr>
<tr>
<td>3</td>
<td>77, F</td>
<td>chronic HA</td>
<td>ND</td>
<td>rt parietal convexity SAH</td>
<td>T2 (FLAIR)</td>
<td>normal</td>
<td>ND</td>
</tr>
<tr>
<td>4</td>
<td>28, F</td>
<td>sudden severe HA, nausea, photophobia, vertigo</td>
<td>lt frontal convexity SAH</td>
<td>rt parietal convexity SAH</td>
<td>T1, FLAIR</td>
<td>normal</td>
<td>ND</td>
</tr>
<tr>
<td>5</td>
<td>30, F</td>
<td>HA, tonic–clonic seizures, AMS, AMS, anisocoracia</td>
<td>rt parietal convexity SAH</td>
<td>lt parietal convexity SAH (FLAIR)</td>
<td>normal</td>
<td>ND</td>
<td>↑WBCs</td>
</tr>
<tr>
<td>6</td>
<td>63, M</td>
<td>AMS, anisocoracia</td>
<td>rt parietal convexity SAH</td>
<td>ND</td>
<td>ND</td>
<td>ND</td>
<td>↑PT/INR, ↑WBCs, anemia</td>
</tr>
<tr>
<td>7</td>
<td>40, M</td>
<td>HA, AMS</td>
<td>rt parietal convexity SAH</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, anemia, ↑ESR</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>64, M</td>
<td>sudden severe HA, AMS</td>
<td>lt frontal, temporal, parietal convexity SAH, HCP</td>
<td>lt cerebral SAH (T1, FLAIR)</td>
<td>narrowing of proximal lt ACA &amp; rt VA</td>
<td>↑WBCs, anemia, ↑D-dimer</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>49, F</td>
<td>lt side weakness, AMS</td>
<td>rt frontal convexity SAH</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, anemia, ↑D-dimer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>33, F</td>
<td>tonic–clonic seizures, HA</td>
<td>rt frontal SAH, herniation</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, anemia, ↑D-dimer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>69, F</td>
<td>rt arm weakness</td>
<td>normal</td>
<td>rt frontal convexity SAH</td>
<td>T2 (STAR)</td>
<td>normal</td>
<td>↑WBCs, anemia</td>
</tr>
<tr>
<td>12</td>
<td>55, F</td>
<td>sudden severe HA, rt leg weakness</td>
<td>bilat frontal convexity SAH</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, anemia, ↑D-dimer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>52, F</td>
<td>sudden severe HA, rt homonymous hemianopia, AMS</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, ↑WBCs, ↑protein</td>
<td>antibody screen for vasculitis negative, ↑WBCs, anemia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>14</td>
<td>46, M</td>
<td>AMS, anisocoracia</td>
<td>rt parietal convexity SAH</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, anemia, ↑D-dimer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>15</td>
<td>79, F</td>
<td>seizures</td>
<td>rt frontotemporal SAH</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, anemia, ↑D-dimer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>16</td>
<td>35, F</td>
<td>sudden severe HA, photophobia, lt numbness &amp; weakness</td>
<td>rt parietal convexity SAH</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, anemia, ↑D-dimer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>17</td>
<td>33, F</td>
<td>gradual-onset severe HA</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, ↑WBCs, ↑protein</td>
<td>antibody screen for vasculitis negative, ↑WBCs, anemia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>18</td>
<td>27, F</td>
<td>daily severe HA</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, ↑WBCs, ↑protein</td>
<td>antibody screen for vasculitis negative, ↑WBCs, anemia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>19</td>
<td>86, M</td>
<td>chronic HA</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, anemia, ↑D-dimer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>25, F</td>
<td>HA, rt weakness, vomiting</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, xanthochromia</td>
<td>↑WBCs, anemia, ↑D-dimer</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* ACA = anterior cerebral artery; AMS = altered mental status; DSA = DS angiography; ESR = erythrocyte sedimentation rate; HA = headache; HCP = hydrocephalus; IgG = immunoglobulin G; MCA = middle cerebral artery; MRA = MR angiography; ND = not done; PT/INR = prothrombin time/international normalized ratio; RBCs = red blood cells; SCA = superior cerebellar artery; STAR = T2* MR imaging sequence; VA = vertebral artery; WBCs = white blood cells; ↑ = elevated.
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and that of Spitzer et al. (42%), which is greater than that reported for aneurysmal SAH.43

Several explanations for these differences in presentation may exist. Spitzer and colleagues40 suggested that the lower incidence of thunderclap headache and nuchal rigidity may reflect a smaller volume of subarachnoid blood, its confinement outside the basal cisterns, or a nonarterial source of hemorrhage. It may also relate to inherent differences in the causes of the disease between the 2 populations of patients with SAH. Whereas SAH from a ruptured aneurysm is an isolated event that accounts for the sudden and severe appearance of symptoms, many of the disorders leading to isolated convexity SAH (for example, PRES, lupus vasculitis, and cerebral venous thrombosis) cause headaches in the absence of SAH. It is therefore not surprising that headaches associated with convexity SAH have a variable rapidity of onset as well as in overall quality and severity. The higher incidence of seizures and focal neurological deficits associated with convexity SAH probably reflects the more extensive parenchymal involvement of many of the underlying causes. Local cortical irritation by subarachnoid blood may also be a contributing factor.40

Radiological Evaluation

A second observation in this study is the need for a systematic and often multimodal approach toward identifying underlying causative disorders. Head CT scanning without contrast agents continues to be the study of choice for screening acute cerebral hemorrhage. In our series, head CT scanning was diagnostic of convexity SAH in 18 (95%) of the 19 patients initially examined. One patient (Case 11) had normal CT findings, but subsequent MR imaging revealed subacute blood over the right frontal

### TABLE 2
Overview of origins, risk factors, treatments, and outcomes in 20 patients with isolated convexity SAH*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>Origin</th>
<th>Risk Factors</th>
<th>Treatment</th>
<th>Outcome at Time of Discharge</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>18, F</td>
<td>lupus vasculitis</td>
<td>systemic lupus erythematosus, HTN</td>
<td>antihypertensives, pulse steroid therapy</td>
<td>neurological status at baseline, no further seizures</td>
</tr>
<tr>
<td>2</td>
<td>74, F</td>
<td>unknown</td>
<td>HTN</td>
<td>nimodipine, Keppra, antihypertensives, analgesics</td>
<td>neurological status at baseline, HA well controlled</td>
</tr>
<tr>
<td>3</td>
<td>77, F</td>
<td>unknown</td>
<td>diabetes</td>
<td>acetaminophen</td>
<td>neurological status at baseline, HA well controlled</td>
</tr>
<tr>
<td>4</td>
<td>28, F</td>
<td>postpartum PRES</td>
<td>none</td>
<td>Keppra, acetaminophen, antihypertensives</td>
<td>neurological status at baseline, no further seizures</td>
</tr>
<tr>
<td>5</td>
<td>30, F</td>
<td>postpartum PRES</td>
<td>HTN</td>
<td>Dilantin, nimodipine, diazepam, antihypertensives</td>
<td>neurological status at baseline, no further seizures</td>
</tr>
<tr>
<td>6</td>
<td>63, M</td>
<td>anticoagulation therapy</td>
<td>sepsis, lower-extremity amputation, atrial fibrillation due to end-stage liver disease, alcoholism, diabetes</td>
<td>Dilantin, steroids, cessation of warfarin platelet transfusion</td>
<td>hospital course complicated by vasospasm &amp; HCP; neurological status improved to near baseline (mild cognitive deficits)</td>
</tr>
<tr>
<td>7</td>
<td>40, M</td>
<td>thrombocytopenia</td>
<td>deep venous thrombosis</td>
<td>Keppra, nimodipine, IVC filter, hyperdynamic therapy, ventriculostomy, analgesics</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>8</td>
<td>64, M</td>
<td>unknown</td>
<td>deep venous thrombosis</td>
<td>Keppra, nimodipine, IVC filter, hyperdynamic therapy, ventriculostomy, analgesics</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>9</td>
<td>49, F</td>
<td>hypertensive microangiopathy</td>
<td>HTN</td>
<td>antihypertensives, analgesics</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>10</td>
<td>33, F</td>
<td>immunosuppressive PRES (post lung transplant)</td>
<td>cyclosporin treatment</td>
<td>Dilantin, Depakote</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>11</td>
<td>69, F</td>
<td>unknown</td>
<td>none</td>
<td>analgesics</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>12</td>
<td>55, F</td>
<td>unknown</td>
<td>none</td>
<td>acetaminophen</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>13</td>
<td>52, F</td>
<td>Call–Fleming syndrome</td>
<td>HTN, diabetes</td>
<td>steroids, Topamax, analgesics, brain biopsy</td>
<td>neurological status at baseline, HA resolved</td>
</tr>
<tr>
<td>14</td>
<td>46, M</td>
<td>immunosuppressive PRES (post stem cell transplant)</td>
<td>leukemia</td>
<td>none</td>
<td>neurological status at baseline, HA resolved</td>
</tr>
<tr>
<td>15</td>
<td>79, F</td>
<td>thrombocytopenia &amp; anticoagulation therapy</td>
<td>anticoagulation therapy for mechanical heart valve</td>
<td>cessation of warfarin</td>
<td>neurological status at baseline, HA resolved</td>
</tr>
<tr>
<td>16</td>
<td>35, F</td>
<td>unknown</td>
<td>HTN</td>
<td>Dilantin, acetaminophen nitroglycerin, ibuprofen</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>17</td>
<td>33, F</td>
<td>postpartum cerebral angioaccessory</td>
<td>HTN peripartum</td>
<td>Dilantin, acetaminophen nitroglycerin, ibuprofen</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>18</td>
<td>27, F</td>
<td>postpartum PRES</td>
<td>HTN, amphetamine use</td>
<td>nimodipine, Solu-Medrol, prednisone, Topamax, antihypertensives, ibuprofen</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>19</td>
<td>86, M</td>
<td>unknown</td>
<td>none</td>
<td>analgesics</td>
<td>neurological status at baseline</td>
</tr>
<tr>
<td>20</td>
<td>25, F</td>
<td>drug-induced vasculopathy</td>
<td>dietary supplement (ephedra)</td>
<td>Dilantin, analgesics, cessation of dietary supplement</td>
<td>neurological status at baseline</td>
</tr>
</tbody>
</table>

* HTN = hypertension; IPH = intraparenchymal hemorrhage; IVC = inferior vena cava.
convexity. The sensitivity of CT scanning declines markedly in the subacute setting. We therefore recommend MR imaging (which is clearly superior to CT by Day 5), as the initial neuroimaging study when patients present in delayed fashion.

**Differential Diagnosis and Workup**

Following initial diagnosis of convexity SAH, further evaluation is required to identify a potential underlying cause and a direct therapy specific to the origin of the SAH (for example, antihypertensive therapy for PRES, platelet transfusion for thrombocytopenia, antiocoagulation for venous thrombosis, and steroids for vasculitis). Because the differential diagnosis is broad, subsequent workup must evaluate for the varied causes. Vital signs should be obtained in all patients, because hypertension is a key feature of PRES (it was the cause in 5 cases in our series). All patients should be tested for the platelet count and prothrombin time to rule out thrombocytopenia or coagulopathy (the cause in 3 cases in our series).

In the absence of clinical or laboratory clues to suggest a definitive origin, we recommend that patients undergo MR imaging, which is sensitive to many causes of convexity SAH including PRES,4 mass lesions,40 vasculitis,2 dural sinus thrombosis,6,27 cortical venous thrombosis,4,28 cerebral AVMs,12 dural AVFs,44 and cavernous malformations.15 In patients with unrevealing MR imaging studies, DS angiography should be considered, because it can demonstrate narrowing of medium-sized arteries (of concern as a sign of vasculitis or vasculopathy), segmental cerebral vasoconstriction (a sign of Call–Fleming syndrome), and aneurysms or other vascular malformations. In our series, DS angiography led to the diagnosis of a specific vasculopathy in 2 patients; vasculitis in one, and Call–Fleming syndrome in the other.

Although not diagnostic, CSF analysis in the appropriate clinical context can provide corroborative information in support of a specific diagnosis. For example, pleocytosis, elevated protein, and increased immunoglobulin G suggest vasculitis (the cause of 1 case in our series). Additional hematological studies, including erythrocyte sedimentation rate, C-reactive protein, and serum antibody screens can often be helpful.

Using the aforementioned diagnostic schema, we identified a definitive underlying cause in 13 of 20 patients. The most common cause was PRES (5 patients, 25%), which is a clinicoradiological entity characterized by acute to subacute onset of headache, seizures, altered mental status, vomiting, and visual symptoms ranging from blurred vision to cortical blindness. On radiological evaluation, white and gray matter edema is typically seen in the parietooccipital lobes (Fig. 1), although more anterior involvement may occur. Posterior reversible encephalopathy syndrome may result from uncontrolled hypertension, immunosuppression, eclampsia, renal failure, drug overdose, connective tissue disorders, acute intermittent porphyria, and hypercalcemia.13,15,20,21,37,38,40,41 The syndrome is transient if treated early, but delayed diagnosis may lead to permanent neurological deficits.1 Four of our 5 patients with PRES had favorable outcomes, but 1 developed severe cerebral edema, resulting in herniation and brain death. Our experience suggests that convexity SAH on initial head CT scans may be the first radiographic clue to PRES. Because MR imaging is more sensitive in demonstrating the associated and often diagnostic parenchymal changes, this imaging modality should be performed in any patient in whom PRES is suspected.

Thrombocytopenia or anticoagulant-induced coagulopathy was the identified underlying cause in 3 of our patients. Coagulopathy and thrombocytopenia are known causes of spontaneous SAH in the setting of multisystemic disease.33,36 Management includes cessation of the offending medication, platelet transfusions, fresh-frozen plasma transfusions, and the administration of vitamin K. In the setting of cerebral hemorrhage, therapeutic goals generally include maintaining platelet counts of > 100,000/µL and normalizing the patient’s international normalized ratio.

Angiographically evident vasculopathy was noted in 2 of our cases. One patient had postpartum cerebral angiopathy, a known cause of convexity SAH.42 These patients are usually healthy young women who present with acute headache, seizures, neurological deficits, and angiographic findings consistent with vasospasm or vasculitis.42 Drug-
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induced vasculopathy and SAH have previously been attributed to the dietary supplement ephedra.\textsuperscript{11} Our case represents the second one in which the temporal association between ephedra consumption, isolated SAH, and angiographic abnormalities suggests a causal relationship. Other dietary supplements such as phenylpropanolamine have also been associated with intracerebral hemorrhage.\textsuperscript{10,16}

Lupus vasculitis was identified in 1 of our patients. Subarachnoid hemorrhage in patients with systemic lupus erythematosus is well described.\textsuperscript{18,20} Central nervous system vasculitis may be a secondary manifestation of an underlying systemic vasculitis (for example, Wegener granulomatosis, polyarteritis nodosa, or Churg–Strauss syndrome), or connective tissue disorder (for example, systemic lupus erythematosus).\textsuperscript{20} Alternatively, central nervous system vasculitis may represent a primary condition known as isolated angiitis of the central nervous system.\textsuperscript{19,20,39} Both DS angiography (assessing for narrowing of medium-sized arteries) and MR imaging (assessing for associated parenchymal abnormalities) should be pursued when the diagnosis of vasculitis is being considered. Additional studies such as serum antibody and CSF analyses can often lead to a definitive diagnosis. However, in some circumstances when diagnosis proves elusive, an open brain biopsy may be required.\textsuperscript{14}

Another case of convexity SAH in our series was ascribed to hypertensive microangiopathy. In patients with chronic uncontrolled hypertension, microangiopathic changes to the cerebral vasculature can result in intracranial bleeding. Diagnosis is made based on MR imaging results in the context of chronic hypertension, with characteristic microhemorrhages and lacunae in the corticosubcortical region and deep gray matter.\textsuperscript{22}

The final case of this series was attributed to Call–Fleming syndrome.\textsuperscript{4} This syndrome is characterized by thunderclap headache and reversible cerebral vasoconstriction. The vasoconstriction may last days or weeks and cause infarction. It has been associated with the postpartum period, migraines, use of cocaine or amphetamines, and the abrupt cessation of selective serotonergic receptor inhibitors.\textsuperscript{26} In the present study we report on a 52-year-old woman presenting with thunderclap headache and right homonymous hemianopia. Initial head CT studies demonstrated SAH in the bilateral parietal lobes, and brain MR imaging revealed bilateral parietooccipital SAH. The initial DS angiography study was negative for vascular abnormality. A second session of DS angiography performed at 7 days showed subtle new narrowing of the proximal SCAs bilaterally (Fig. 2A). Approximately 10 days later, the patient developed new left upper-extremity weakness and worsening visual complaints. The MR imaging demonstrated small acute infarctions primarily in the right parietooccipital convexity and an associated left occipital lobe hematoma. The patient was then taken to the operating room, where clot evacuation and parietal brain biopsy were accomplished. The biopsy was inconclusive for vasculitis but did demonstrate thickening of cortical blood vessels. Repeated angiography showed diffuse segmental narrowing of bilateral internal carotid and verteobasilar arteries (Fig. 2B). She was treated with calcium channel blockers and ultimately discharged to inpatient rehabilitation with improved left upper-extremity strength and vision. A final angiogram obtained 2 weeks later showed complete resolution of the vasculopathy. Because the patient’s initial presentation resembled that described by Call et al.,\textsuperscript{4} and subsequent DS angiography showed segmental vasoconstriction, we considered her condition to be Call–Fleming syndrome. It is the first such case to be associated with SAH.

Other possible causes of convexity SAH reported in the literature, but not seen in our case series, include mycotic aneurysm,\textsuperscript{23,43} superficial AVM,\textsuperscript{44} dural AVF,\textsuperscript{44} transmural arterial dissection,\textsuperscript{45} cavernous malformation,\textsuperscript{40,45} dural sinus/cortical vein thrombosis,\textsuperscript{4,27,28} brain abscess,\textsuperscript{32} and brain neoplasm.\textsuperscript{40} All of these potential causes of spontaneous isolated convexity SAH should be pursued with rigorous diagnostic workup, as detailed earlier.

![Fig. 2. Case 13. Angiograms demonstrating convexity SAH associated with Call–Fleming syndrome. A: Right vertebral angiogram, anteroposterior injection. Subtle mild irregularity of both SCAs is present, a change from the initial study obtained on admission. B: A third angiogram obtained 10 days later, following clinical deterioration and an MR imaging study showing multiple small acute infarcts in the right parietooccipital convexity with an associated left occipital hematoma (MR image not shown). The cerebral angiogram depicted multiple new severe arterial narrowing, which were worst in the posterior circulation, but also involved both intracranial carotid artery territories.](image-url)
Clinical Course

A final observation involves the generally benign clinical course of patients with isolated convexity SAH, compared with aneurysmal SAH. The majority of patients in our series (75%) had complete or near-complete return to their premorbid condition by the time of discharge (mean hospital stay 16.5 days). The clinical course, however, was highly dependent on the underlying origin of the disease. For example, all patients without an underlying causative disorder had a favorable outcome, whereas only 8 of 13 patients with a definitive underlying diagnosis fared as well. Hydrocephalus and symptomatic cerebral vasospasm were rare, with only 1 patient (who experienced the largest volume of subarachnoid blood in our series) developing such complications, neither of which led to long-term clinical sequelae.

Conclusions

Spontaneous isolated SAH over the cerebral convexity is rare. Compared with aneurysmal SAH, patients present less commonly with thunderclap headache and nuchal rigidity and more commonly with seizures and focal neurological deficits. Spontaneous convexity SAH carries a broad differential diagnosis. The most common cause in our series was PRES, which accounted for 22% of cases. Diagnostic workup should include head CT scans obtained without contrast agent as well as platelet count and prothrombin time. Most patients also require brain MR imaging, and selected individuals may benefit from diagnostic catheter angiography. A CSF evaluation and certain hematological studies are indicated in the case of suspected vasculitis. General treatment measures include serial neurological evaluations, seizure prophylaxis, and analgesics. Prophylactic use of nimodipine may be reserved for those patients with the largest volume of subarachnoid blood. Treatment directed toward the underlying condition (when identified) is paramount. Finally, although complications specific to the underlying disorder can occur, the overall prognosis for this patient population is generally favorable.

Disclaimer

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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