

Pathologically Confirmed Cryptic Vascular Malformation as a Cause of Convexity Subarachnoid Hemorrhage: Case Report

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BACKGROUND AND IMPORTANCE: We report a rare case of pathologically confirmed cryptic vascular malformation as a cause of primary convexity subarachnoid hemorrhage (SAH) of unknown etiology.

CLINICAL PRESENTATION: A 48-year-old woman presented with sudden severe headache. Localized right convexity SAH was observed on computed tomography (CT) scan, but the origin could not be detected despite extensive workup covering the entire head by using 3.0-Tesla magnetic resonance (MR) imaging with MR angiography and CT angiography combined with venous-phase imaging with a 320-detector row CT scanner. Subsequent digital subtraction angiography (DSA) performed 2.5 hours after admission failed to reveal any cause of SAH; however, a right frontoparietal avascular region was suspected to be due to a newly developed intracerebral hematoma. The lesion was simultaneously confirmed by angiographic cone-beam CT imaging. Because she remained neurologically intact, we decided to perform a follow-up study later with medical management. However, she developed left hemiparesis 3 hours after DSA. CT scan demonstrated progression of the hematoma, and her symptoms gradually worsened. Emergent surgical exploration along the SAH superficial to the postcentral sulcus and hematoma evacuation were performed, with favorable functional outcome. Pathological examination confirmed cryptic vascular malformation with several abnormally dilated arterioles within the subarachnoid space surrounded by a thick SAH clot.

CONCLUSION: It is important to consider the possibility of ruptured cryptic vascular malformation as a cause of nontraumatic nonaneurysmal convexity SAH when recurrent hemorrhage occurs despite thorough diagnostic workup, because surgical resection may be the only curative treatment option to eliminate the risk of rebleeding and disabling symptoms.

KEY WORDS: Convexity subarachnoid hemorrhage, Cryptic vascular malformation, Subarachnoid hemorrhage of unknown etiology

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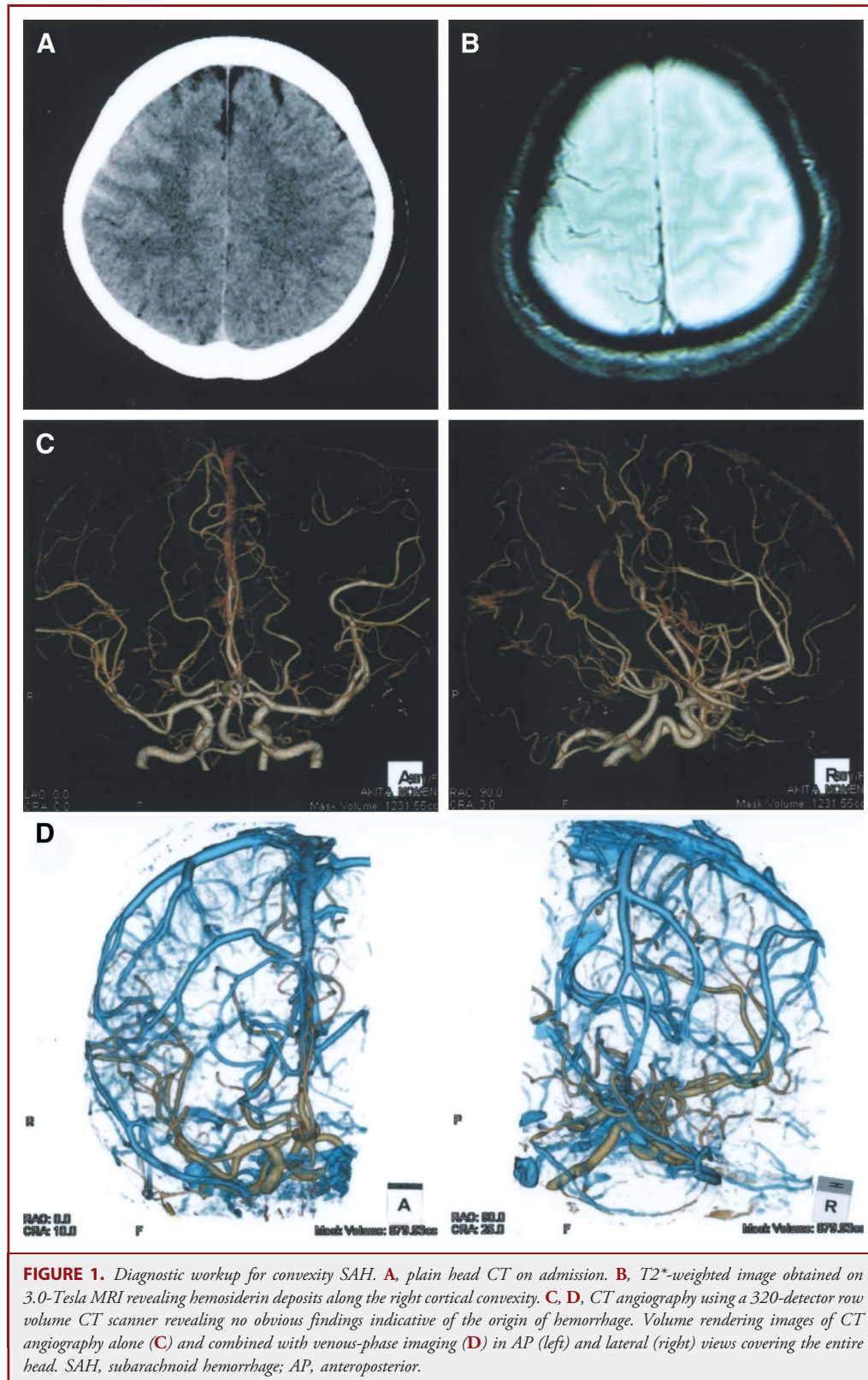
Nontraumatic subarachnoid hemorrhage (SAH) along the cortical convexity is rare and may often be nonaneurysmal in origin and not demonstrable angiographically. Diverse etiologies have been suggested for its occurrence, including reversible cerebral vasoconstriction syndrome, posterior reversible leukoencephalopathy syndrome, cerebral amyloid angiopathy, dural sinus/cortical vein thrombosis,

coagulopathy, cocaine use, lupus vasculitis, cavernoma, and brain abscess.^{1–4} We report another potential cause of primary isolated convexity SAH in an extremely rare case of ruptured cryptic vascular malformation in the subarachnoid space in which emergent surgical exploration and resection of its origin effectively eliminated the risk of rebleeding and life-threatening/disabling symptoms.

ABBREVIATIONS: DSA, digital subtraction angiography; ICH, intracerebral hematoma; SAH, subarachnoid hemorrhage

CASE REPORT

A 48-year-old previously healthy woman presented in a neurosurgical clinic with sudden severe headache



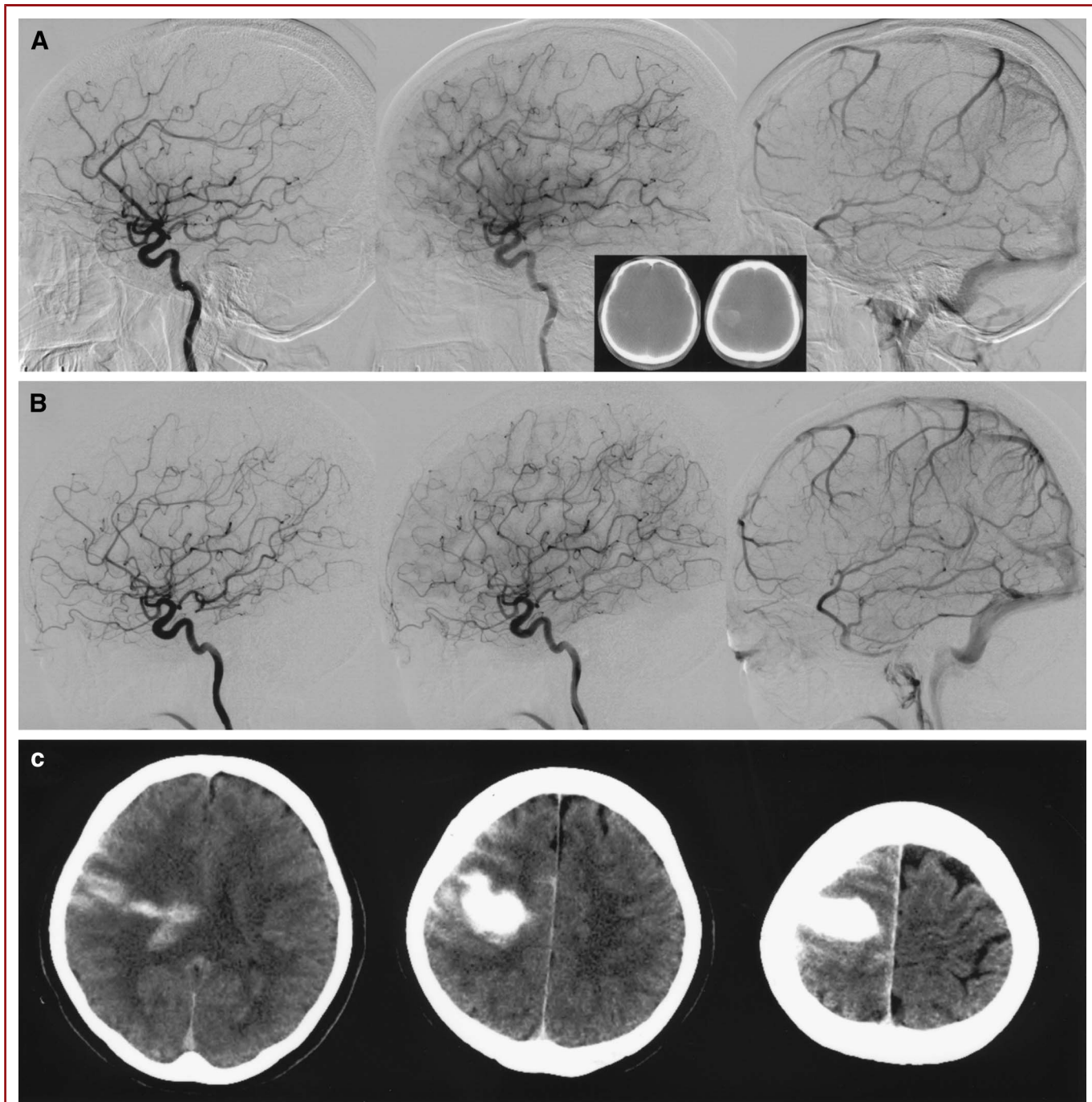


FIGURE 2. **A**, cerebral DSA of the right internal carotid artery in the lateral view performed 2.5 hours after admission shows an avascular region in the frontoparietal lobe without extravasation (middle photograph, early venous phase) suspected to be due to newly developed intracerebral hematoma. The lesion was simultaneously confirmed by digital flat-panel detector angiographic cone-beam CT imaging (**A**, inset). **B**, postoperative DSA performed 1 week after surgery demonstrating the disappearance of this region. **C**, CT scans performed when focal neurological symptoms occurred 3 hours after DSA demonstrated the progression of intracerebral hematoma deep along the convexity SAH protruding into the right lateral ventricle. DSA, digital subtraction angiography; SAH, subarachnoid hemorrhage.

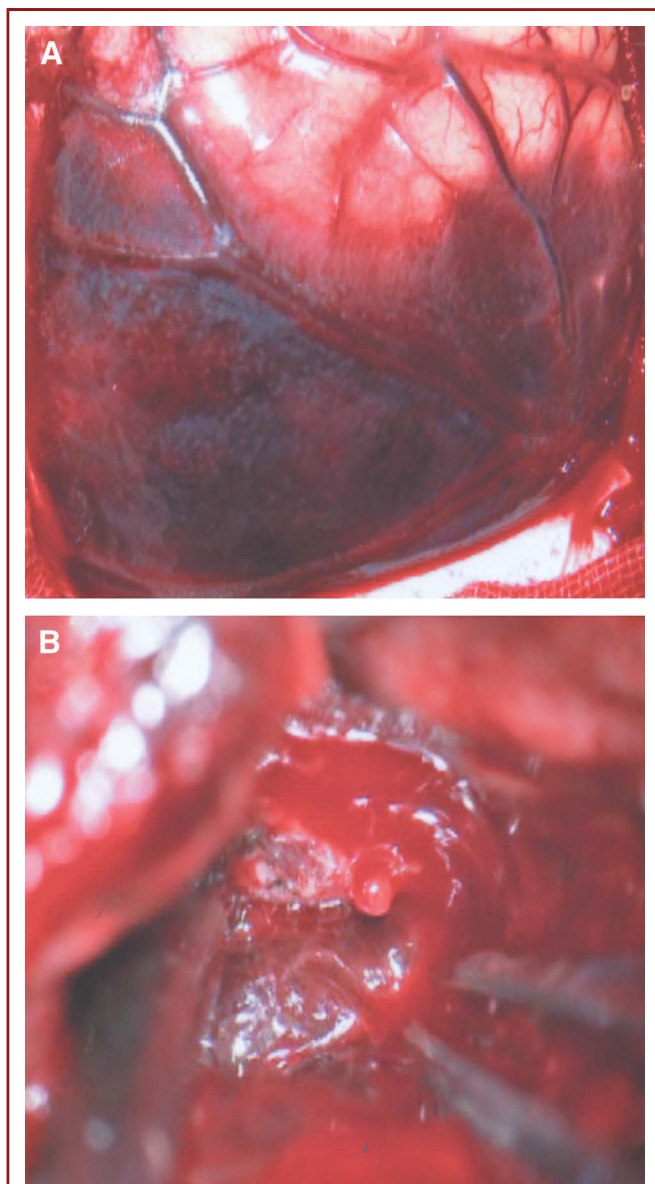


FIGURE 3. Intraoperative photograph. **A**, isolated SAH in the right convexity approached via a temporoparietal craniotomy. **B**, active bleeding from pseudoaneurysms along with an abnormal tangle of dilated subpial vessels superficial to the postcentral gyrus abutting the hematoma cavity. There are no obvious findings of nidus formation or venous drainage in the surgical field. SAH, subarachnoid hemorrhage.

preceded by transient numbness of the right hand. She was neurologically intact on examination, but localized right convexity SAH was observed on plain head computed tomography (CT) scan (Figure 1A). She was then referred to the stroke service of our center for further diagnosis and treatment. Despite extensive radiological workup covering the entire head using 3.0-Tesla brain magnetic resonance (MR) imaging (Figure 1B) with MR angiography (MAGNETOM Verio,

Siemens Medical Solutions, Erlangen, Germany) and CT angiography combined with venous-phase imaging with a 320-detector row volume CT scanner (Aquilion ONE, Toshiba Medical Systems, Tokyo, Japan) covering the entire head (Figure 1C and 1D), no abnormal findings indicative of the cause of hemorrhage were noted throughout the arterial and venous phases.

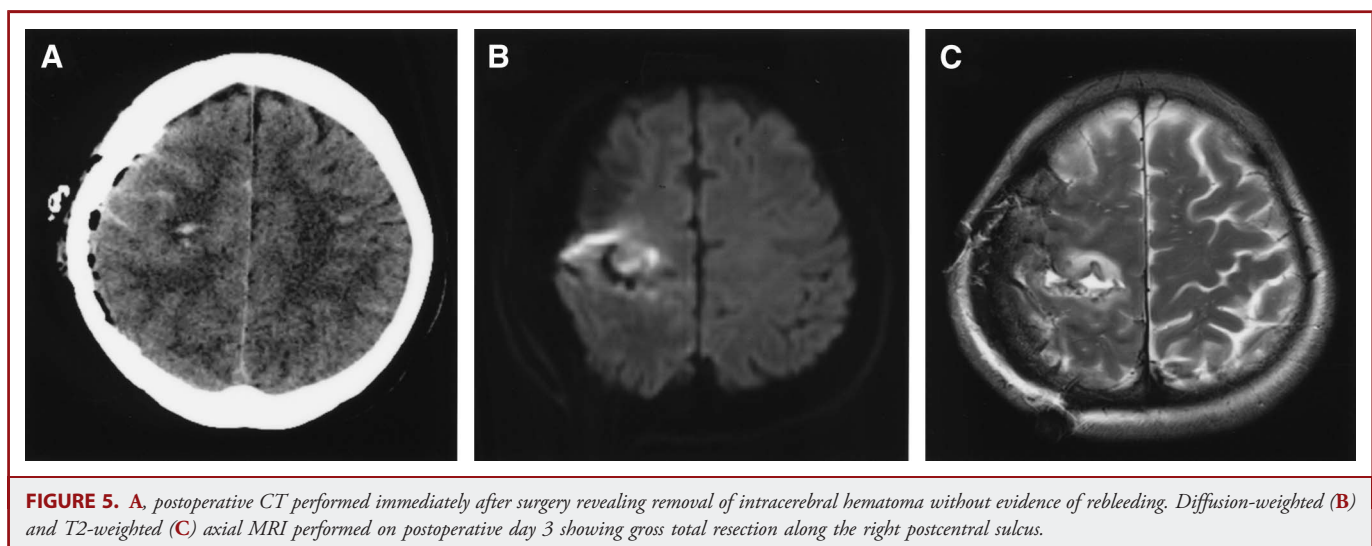
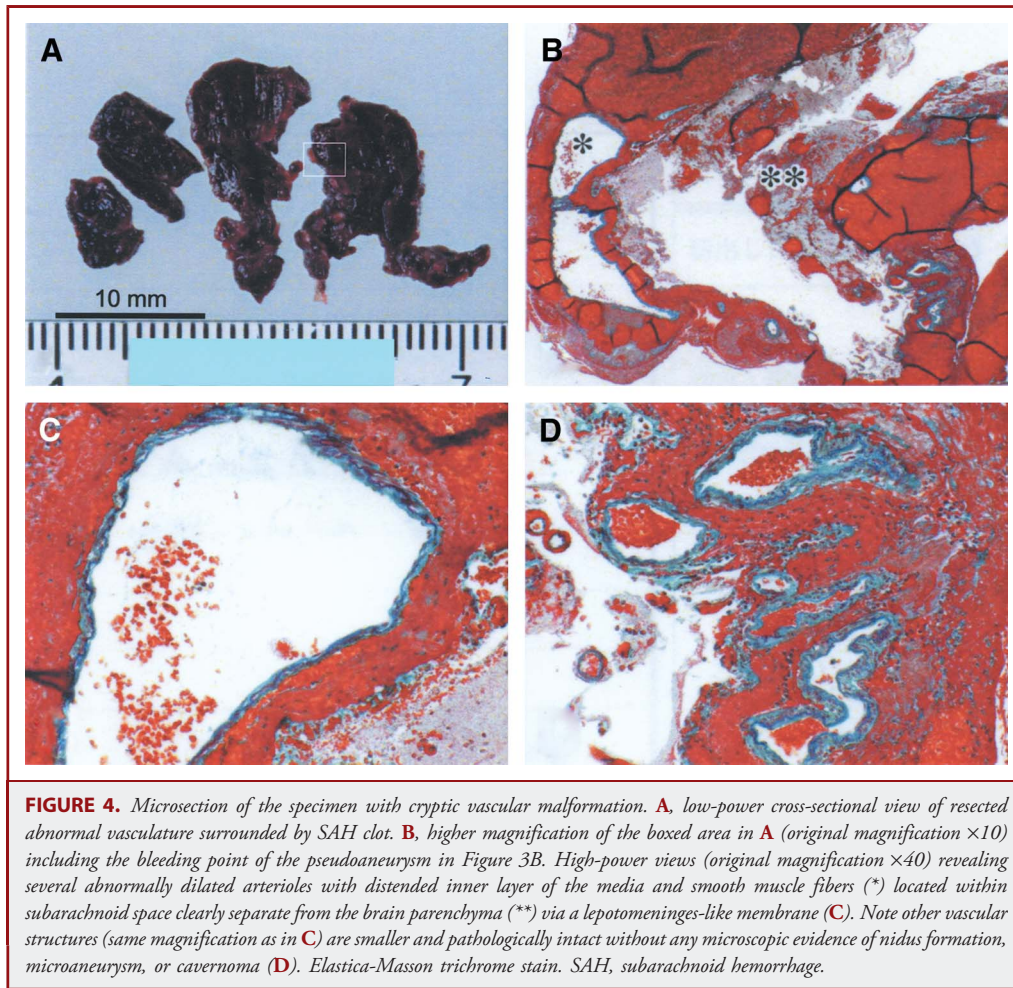
Subsequent digital subtraction angiography (DSA) with a biplanar digital flat-panel detector angiography system (AXIOM Artis dBA, Siemens Medical Solutions, Erlangen, Germany) performed 2.5 hours after admission under mild sedation for further diagnostic yield failed to reveal any cause of SAH including tumor blush, multifocal vasospasm, or vascular malformation, although a right frontoparietal avascular region without extravasation (Figure 2A) was suspected to be due to a newly developed intracerebral hematoma (ICH). The lesion was simultaneously confirmed by flat-panel detector angiographic cone-beam CT scan imaging (Syngo Dyna-CT, Siemens Medical Solutions, Erlangen, Germany) (Figure 2A, inset).

Because she remained neurologically intact, we decided to perform medical management (systolic blood pressure reduction <140 mm Hg) to avoid hematoma expansion⁵ and to perform a follow-up study later after reduction of the mass effect. However, she developed left hemiparesis and numbness of the left hand accompanied by nausea and vomiting 3 hours after DSA. CT scan demonstrated progression of intracerebral hematoma deep along the convexity SAH protruding into the right lateral ventricle (Figure 2C). The symptoms progressively worsened, and left hemispatial neglect developed within the next hour.

In view of the negative workup and accessible location with progressive focal neurological deficits, we elected to perform emergent surgical exploration of the convexity SAH with ICH via a temporoparietal craniotomy 2 hours after the initial deterioration (Figure 3A). Intraoperatively, active bleeding from an abnormal cluster of subpial vessels superficial to the postcentral sulcus abutting the hematoma cavity was seen (Figure 3B). No arterialized cortical veins, microaneurysm, cavernous angioma, or hemorrhagic tumor was observed. The lesion was resected en bloc following hematoma evacuation. Pathological examination confirmed cryptic vascular malformation with several abnormally dilated arterioles with a distended inner layer of the media and smooth muscle fibers within the subarachnoid space surrounded by a thick SAH clot (Figure 4). Postoperative CT revealed removal of intracerebral hematoma without evidence of rebleeding (Figure 5A). MRI revealed gross total resection with reduction of mass effect in the eloquent area (Figure 5B and 5C). The avascular region disappeared on postoperative DSA performed 1 week after resection (Figure 2B). Further recovery was uneventful, and at the time of discharge (approximately 3 months after surgery) no deficits were present other than mild numbness of the left hand.

DISCUSSION

This is, to the authors' knowledge, the first report of ruptured cryptic vascular malformation presenting with primary isolated convexity SAH of unknown etiology confirmed pathologically by examination of a surgical specimen resected directly from the bleeding point. One cohort² and 2 case series^{3,4} have been published on nontraumatic nonaneurysmal SAH in the cortical convexity in which 2



common patterns of presentation were identified: younger patients ≤ 60 years with sudden onset of headache similar to aneurysm rupture are frequently diagnosed with a primary vasoconstriction syndrome following exclusion of vascular malformations on repeated DSA, and older patients who have temporary sensory or motor symptoms and typically exhibit amyloid angiopathy. These studies consistently indicate a benign clinical course and outcome for convexity SAH in younger patients, but note the possibility of small superficial arteriovenous malformations or microangiomas that may have escaped detection in radiological workup, but that can be lethal.

The presence of acute ICH in a young and healthy adult in the absence of an obvious cause is strongly suggestive of an underlying pathological condition. It should be noted that convexity SAH carries significant risks of recurrent ICH and high mortality (50%) in the elderly, mostly in association with amyloid angiopathy.² On the other hand, the natural history of vascular malformations that cause primary isolated convexity SAH and the actual risk of massive hemorrhage of them are unclear, because fewer than 5% of all ruptured arteriovenous malformations causes extravasation in the subarachnoid space without ICH.⁶ Given the pathophysiological similarity of amyloid angiopathy originating in the subarachnoid space, particularly the cerebral sulci, and causing multiple rupture of meningeal arteries,^{7,8} it may also be the case that isolated vascular malformations located primarily in the subarachnoid space of the convexity (Figure 4) are a source of ICH. Despite the lack of evidence for nidus formation or major draining veins, the presence of pseudoaneurysms, which increase the risk of hemorrhage,⁹ suggests the vulnerability of dilated vessels to hemodynamic stress, triggering vessel rupture.

The term "cryptic" has been proposed by Crawford and Russell to describe *small* vascular malformations less than 2 to 3 cm in diameter that cause spontaneous cerebral hemorrhage.¹⁰ Currently, the combination of meeting the criterion for size (< 1 cm in diameter and often a few millimeters in size)¹¹ and being angiographically occult^{12,13} can be used to define lesions. Despite extensive workup using advanced modalities of high diagnostic yield (3.0-Tesla MRI/MRA, 3D-CT DSA with 320-detector row CT system, and flat-panel detector DSA with volumetric CT imaging system), the lesion in the present case was too small to specify its origin radiologically without evidence of newly developed ICH and/or neurological deterioration and to proceed with early surgical exploration.¹⁴ Of course, we still cannot rule out the possibility of negative catheter DSA findings in situations in which mass effect from the intraparenchymal hemorrhage can compress and stop flow into the microarteriovenous malformation,⁹ preventing it from being visualized on the angiogram, and thus repeated DSA may have contributed to obtaining a useful diagnostic yield. In any event, it was fortunate in this case that intraoperatively we could obtain a specimen of vascular malformation from a few abnormally dilated small vessels that was successfully visualized

in the subarachnoid space by careful inspection of the hematoma cavity and SAH clots and examine it for abnormal vasculature.

The management of angiographically and MRI-negative convexity SAH has not been clearly defined, because the risk of rebleeding is not clearly known. However, based on these findings and those already published in the literature regarding intraparenchymal hemorrhage,¹⁴ surgical exploration of patients strongly suspected to have an underlying origin, if there is no ICH and only convexity SAH, may be considered. In particular, the presence of symptomatic hemorrhage accompanied by progressive and/or recurrent focal signs (eg, transient sensory/motor deficits, as observed in convexity SAH caused by amyloid angiopathy²) in an accessible and noneloquent area is another factor facilitating early/emergent surgical inspection. On the other hand, if the lesion is surgically inaccessible or asymptomatic, waiting and conservative management are possible with follow-up imaging studies for the resolution of SAH clots without recurrence at least during the time of hospital stay (approximately 16.5 days)³ for most patients presenting with a benign clinical course.

In conclusion, it is important to consider the possibility of ruptured cryptic vascular malformation as a cause of nontraumatic nonaneurysmal convexity SAH when recurrent hemorrhage is observed despite rigorous diagnostic workup, because surgical resection may be the only curative treatment option and eliminate the risk of rebleeding and disabling symptoms.

Disclosure

The authors have no personal financial or institutional interest in any of the drugs, materials, or devices described in this article.

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COMMENT

In this case report, the authors describe a patient with a cryptic arteriovenous malformation (AVM) presenting with isolated subarachnoid

hemorrhage (SAH) initially on computed tomography (CT) scan that was angiographically normal, followed by a rebleed, resulting in a symptomatic intraparenchymal hematoma requiring surgical evacuation. During evacuation, the contents of the hematoma were found to contain a small bundle of abnormally dilated vessels consistent with an AVM, later confirmed histologically. The authors are correct in stating that there were no previous reports of a cryptic AVM initially presenting with only SAH. Others have described cryptic lesions identified at the time of surgical exploration for intraparenchymal hemorrhages, and these references are cited and briefly discussed by the authors. Surgical exploration in patients with angiographically and magnetic resonance imaging-negative, convexity SAH without IPH (intraparenchymal hemorrhage), is not a reasonable option for patients. However, cryptic AVMs should be in the differential diagnosis in such cases, particularly because the AVM may not be visualized radiographically as a result of thrombosis or from compression by the mass effect of IPH. In short, it still appears that surgical exploration in such a case should be reserved only for those who also have a symptomatic IPH with mass effect, while those with angiographic- and magnetic resonance imaging-negative convexity SAH alone, should be followed carefully with follow-up imaging studies.

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