Pearls and Oy-sters: Small but consequential
Intracerebral hemorrhage caused by lenticulostriate artery aneurysm

PEARLS

- Cerebrovascular anomalies such as aneurysms and arteriovenous malformations (AVMs) are rare but important causes of intracerebral hemorrhage (ICH) and are associated with a significantly higher risk of rebleeding; early detection and treatment are crucial.
- Noninvasive modalities such as CT and magnetic resonance angiography (MRA) are well tolerated, rapid, and widely available but have limited sensitivity, especially for detection of small aneurysms or AVMs.
- It is important to perform diagnostic cerebral angiography in patients with high clinical suspicion of an underlying vascular malformation or aneurysm even if initial screening CT angiography or MRA is negative.
- In cases of suspected aneurysmal rupture in young patients at high risk without predisposing factors, a repeat conventional angiogram may be necessary in making the diagnosis.

OY-STERS

- Patients with ICH who are young, normotensive, and who have hematomas of unusual shape, location, or with marked subarachnoid hemorrhage (SAH), are at high risk for an underlying cerebrovascular anomaly and should have extensive vascular evaluation.

CASE REPORT

A 41-year-old, right-handed woman presented to the hospital with sudden onset of left-sided weakness. She had a history of borderline hyperlipidemia and a herniated disk treated with laminectomy and fusion with resultant chronic pain treated with an intrathecal pump. Earlier in the day, she had visited the outpatient pain clinic for adjustment of intrathecal medications. Hours later, she started to experience episodes of left leg shaking followed by generalized left-sided weakness. On the way to the hospital, she developed headache, diaphoresis, and agitation.

The patient’s blood pressure was 125/88 mm Hg on arrival to the emergency department. She had a right gaze preference, left-sided visual-spatial and limb asomatognosia, and left face, arm, and leg weakness. Her baseline systolic blood pressures at clinic visits were approximately 90 mm Hg. Her blood work did not reveal any thrombocytopenia or coagulopathy nor was she taking any anticoagulants or antiplatelet agents. She had no personal or family history of bleeding diathesis.

Cerebral CT showed an acute ICH measuring 28 mL in volume involving the right insula, superior temporal lobe, and centrum semiovale with mild right to left midline shift and effacement of the right lateral ventricle without hydrocephalus (figure, A). CT angiography showed a patent circle of Willis without clear aneurysm, AVM, or other vascular anomaly. There was a small focal, nonspecific outpouching seen in the right lenticulostriate vessel, which was not close to the epicenter of the hemorrhage (figure, B). She subsequently underwent conventional cerebral angiography 17 hours later, which did not demonstrate any aneurysm, AVM, or arteriovenous fistula. MRI showed no evidence of cerebral cavernomas or AVMs. Over the next 2 days, she had serial cerebral CT scans that demonstrated stability of the hemorrhage. She never had elevated blood pressures.

Four days later, the patient had acute worsening of her headache, decreased consciousness, and worsened left-sided weakness. Repeat cerebral CT showed increased hematoma size from 28 to 38 mL along with intraventricular expansion of the hematoma (figure, C). Repeat cerebral angiography was performed, this time demonstrating the presence of a 3-mm right lateral lenticulostriate branch aneurysm 15 mm distal to the M1 origin adjacent to the intracranial hematoma (figure, D).

This aneurysm was not amenable to endovascular intervention. The patient subsequently underwent a right frontotemporal craniotomy with microsurgical clipping and sacrifice of the lenticulostriate perforator artery proximal to the aneurysm. Postoperative diagnostic cerebral angiography showed successful obliteration of this aneurysm. The patient did not develop any ischemic infarct from clipping of the lenticulostriate perforator artery.

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artery. On postoperative day 2, repeat cerebral angiogram showed secondary vasospasm in the bilateral A1 segments, which was treated with intra-arterial verapamil. She was also treated with a 21-day course of oral nimodipine to reduce the risk of delayed cerebral ischemia.

The patient had a long course of inpatient rehabilitation followed by intensive outpatient physical therapy. Three months later, a follow-up cerebral angiogram showed no residual aneurysm. One year later, she still had difficulties using her left arm secondary to severe weakness (0/5 strength) in her wrist extensors and finger extensors but was able to ambulate with a left ankle-foot orthotic device.

DISCUSSION This patient presented with an ICH that was caused by rupture of a lenticulostriate artery aneurysm. These aneurysms are rare, and only 23 previous cases have been reported in the literature. Unlike many other aneurysmal hemorrhages, which present with SAH, these aneurysms tend to cause intraparenchymal hemorrhages when ruptured, particularly in or near the basal ganglia. These cases can easily be missed because the appearance and location may lead to a diagnosis of ICH secondary to hypertension. The latter assumption can be misleading; for instance, in one large case series, even in patients who had a history of hypertension, 36% of 144 patients who came to autopsy had a specific structural lesion that had led to ICH.1

One of the most important and urgent management tasks for every ICH is to rule out underlying structural vascular abnormalities such as AVMs and aneurysms. These lesions have high risk of rebleeding, which is associated with high morbidity and mortality, and urgent intervention may be necessary to minimize rebleeding risks. In aneurysmal SAH, patients who rebled in the first 2 weeks had an 80% mortality rate as compared with 41% in those who did not rebled.5 Aneurysmal rebleeding leads to a 12-fold reduction in patients’ chance of survival with functional independence.6 Although our patient had predominantly deep ICH with small SAH, the presence of aneurysm puts her in a high-risk category for rebleeding; in fact, her ICH expansion was likely due to her ruptured aneurysm.

In recent years, CT angiography has emerged as a well-tolerated, noninvasive, rapid, and widely available diagnostic test for the detection of underlying structural abnormalities in ICH. For these reasons, CT angiography is now frequency performed before conventional angiography. Recent literature shows that CT angiography has high accuracy in detecting intracranial aneurysm larger than 4 mm in size (92%–100% sensitivity); however, even with higher-resolution multidetector CT scanners, the sensitivity for smaller aneurysms less than 3 to 4 mm is lower, in the range of 74% to 92%.7 In this patient, CT angiography was not definitive in making a diagnosis of aneurysmal rupture. MRA is another noninvasive technique, but because of lower spatial resolution, its accuracy is limited for aneurysms less than 5 mm.7

Further complicating the diagnosis and treatment in this challenging case was the initially negative conventional cerebral angiography. False-negative cerebral angiography for aneurysm detection can occur if the ruptured aneurysm is thrombosed or if the feeding vessel develops vasospasm. In one case series of patients with ICH, a second angiogram obtained in 22 patients who had initial negative angiography on admission demonstrated 4 underlying vascular malformations.8 In this same group, patients younger than 40 years without history of hypertension were more likely to have identification of occult vascular lesions on repeat testing.6 Overall, a first cerebral angiogram will detect a cerebral aneurysm in 75% to 80% of patients with SAH, with 1% to 2% more aneurysms identified with 1 repeat angiogram.9

The factors favoring more extensive neuroimaging that led to the diagnosis of this patient’s lenticulostriate artery aneurysm included her age (younger than 45 years), lack of comorbid conditions such as hypertension, perisylvian location of the hemorrhage, and expansion of the hemorrhage on hospital day 4. Another presentation that is associated with high likelihood of underlying vascular anomaly is isolated intraventricular hemorrhage.10

Figure Cerebral CT, CT angiography, and cerebral angiography

(A) Initial head CT on presentation with acute intracerebral hemorrhage. (B) CT angiography showing a suspected outpouching near the edge of the hematoma (white arrow). (C) Repeat head CT showing expansion of the hemorrhage. (D) Cerebral angiography showing 3-mm right lateral lenticulostriate branch aneurysm (white arrow).
This patient ultimately underwent clipping of her underlying distal lenticulostriate artery. Similar to previously reported cases in the literature, this patient was treated surgically with a good clinical outcome. In one of the largest case series of 6 patients, 5 patients did well after intervention with low disability scales.\(^8\) This patient’s visual deficits, asomatognosia, and sensory deficits all eventually resolved. Clinically, her strength increased and, over time, she was able to walk and regain functional independence with some residual motor deficits. The generally favorable outcome in these cases makes it even more important that we correctly diagnose and treat the underlying aneurysm before significant rebleeding occurs.

This case illustrates the important consideration that in ICH an underlying treatable structural abnormality should be carefully ruled out particularly if the patient does not carry traditional risk factors for ICH. Furthermore, cerebrovascular anomalies producing ICH may be small (<5 mm) and can be missed on initial angiogram. In patients who are young, are without clear predisposing conditions such as hypertension, and particularly in those who rebleed or have expansion of their bleed, the likelihood of occult vascular malformation or aneurysm is very high. Repeating cerebral angiography in these patients may be necessary to making a lifesaving diagnosis.

**AUTHOR CONTRIBUTIONS**

Concept and design: Drs. Cai, Han, Feske, and Chou. Drafting of the manuscript: Dr. Cai. Critical revision of the manuscript: Drs. Cai, Feske, and Chou. Supervision: Dr. Chou.

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