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Case Report

Intracranial varix of the transverse-sigmoid dural arteriovenous fistula mimicking a ruptured middle cerebral artery aneurysm: A case report

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ABSTRACT

Background: Hemorrhagic stroke is caused by various vascular abnormalities, such as aneurysms, arteriovenous malformations, and dural arteriovenous fistulas (DAVF). Magnetic resonance angiography (MRA) and three-dimensional computed tomography angiography (3DCTA) are used as efficient initial diagnostic modalities in assessing the etiology of hemorrhagic stroke. We describe the unusual case of a false-positive aneurysm on MRA and 3DCTA.

Case Description: A 65-year-old nonhypertensive woman was brought to our hospital with a sudden onset of headache and left hemiparesis. She also had chemosis in the right eye. CT and magnetic resonance imaging showed an intracerebral hemorrhage in the right temporal lobe. MRA and 3DCTA showed a rounded mass suggestive of an aneurysm arising from the bifurcation of the middle cerebral artery (MCA) and also demonstrated an abnormal tortuous vessel contacting with a rounded mass. Digital subtraction angiography showed a transverse-sigmoid sinus DAVF with a varix in contact with the MCA bifurcation. Hematoma evacuation and venous drainage disconnection through the right frontotemporal craniotomy were performed.

Conclusion: This case is very instructive and clinicians should keep in mind that detailed neurological and radiological examinations are essential in obtaining an accurate diagnosis, especially if the bleeding source is similar in shape and location to common lesions (such as a cerebral aneurysm).

Keywords: Cerebral aneurysm, Dural arteriovenous fistula, Varix

INTRODUCTION

Hemorrhagic stroke, caused by various vascular abnormalities, including aneurysms, arteriovenous malformations, and dural arteriovenous fistulas (DAVF),[2,6,19] sometimes requires surgical intervention. Early identification of these lesions is essential for proper treatment and good outcomes.[14,22]

Digital subtraction angiography (DSA) is the gold standard for diagnosing bleeding sources; however, DSA has the disadvantage of a complication rate reported as about 0.8–2.6%.[4,5] Recently, instead of DSA, noninvasive imaging modalities, such as magnetic resonance angiography (MRA) and three-dimensional computed tomography angiography (3DCTA), are increasingly

utilized as an efficient initial modality in evaluating the etiology of hemorrhagic stroke.[20] A thorough examination of images from magnetic resonance imaging (MRI), 3DCTA, and DSA provides an accurate diagnosis in most cases; however, correct diagnosis can be difficult if the bleeding source is similar in shape and location to common lesions (such as cerebral aneurysms).

We report a case of intracranial varix of transverse-sigmoid dural arteriovenous fistula mimicking a ruptured cerebral aneurysm. We also present a brief literature review related to this case.

CASE DESCRIPTION

A 65-year-old nonhypertensive woman was brought to our hospital with sudden onset of severe headache. On arrival, she complained of severe headaches and nausea with Glasgow Coma Scale score of 13. Neurological examination exhibited left hemiparesis and conjunctival chemosis of the right eye. Motor power was Grade 3 on the Medical Research Council scale in the left upper and lower extremities. According to her family, the chemosis appeared a few months ago without any trauma. CT and MRI showed an intracerebral hemorrhage (ICH) in the right temporal lobe without subarachnoid hemorrhage and a rounded mass near the right Sylvian fissure [Figures 1 and 2]. MRA also showed another rounded mass, suggestive of an aneurysm, arising from the bifurcation of the middle cerebral artery (MCA) [Figure 2]. A tentative diagnosis of a ruptured MCA aneurysm causing ICH was made. We planned a craniotomy involving hematoma evacuation and aneurysm clipping then performed 3DCTA to obtain more information about the aneurysm, revealing contact with abnormal tortuous vessels [Figure 3]. Initial MRI showed a dilated right superior orbital vein (SOV) retrospectively [Figure 2]. Her long-standing chemosis and these radiological findings pointed away from an aneurysm as the source of bleeding and we performed DSA for further investigation.

Figure 1: Computed tomography showing intracerebral hemorrhage in the right temporal lobe and the aneurysmal rounded mass (arrow).

DSA demonstrated a transverse-sigmoid junction DAVF fed by branches of the occipital artery. Early retrograde venous drainage flowed into the straight sinus through the inferior petrosal sinus and internal cerebral veins, forming an intracranial varix. DAVF also regurgitated into the SOV and was thought to be the cause of chemosis. No aneurysm was seen on DSA [Figure 4]. The final diagnosis was ICH due to varix but not aneurysmal rupture.

A right frontotemporal craniotomy was performed for hematoma evacuation and venous drainage disconnection of the varix to prevent rebleeding and brain herniation in the acute phase. After a dural incision, the Sylvian fissure was opened entirely. Then, the arterialized vein and varix were detected on the surface of the temporal lobe. After occluding the vein temporally using a clip, we verified the absence of varix flow and brain swelling using Doppler ultrasound. The hematoma was also successfully evacuated after ligating the vein and varix. Although we planned endovascular embolization of the residual fistula regurgitated into the SOV [Figure 5], her family did not consent to additional treatment because of her poststroke cognitive impairment and dementia, including memory disturbance and unilateral spatial neglect. The postoperative course was uneventful and the left hemiparesis was fully resolved within 3 months.

DISCUSSION

Although DSA is a gold standard for diagnosing the pathophysiology of hemorrhagic stroke, it had been replaced by noninvasive MRI and 3DCTA at the time of initial diagnosis due to its invasiveness. Particularly, in a patient with cerebral aneurysms, 3DCTA and MRA demonstrate high sensitivity and specificity rates that can replace DSA for both detection of this condition and surgical planning.[12]

There are some reports that other bleeding sources, such as DAVF,[1,3,11,13] cavernomas,[18,21] hemangioblastomas,[7,15] arachnoid cyst,[17] and gliomas,[16] were mistaken for an aneurysmal rupture on initial imaging modalities. In addition, venous structures, such as a venous loop, can also be mistaken for aneurysms. Kim *et al*. reported a case of a venous loop crossing over the bifurcation of the left MCA mimicking an aneurysm on 3DCTA and performed craniotomy.[9] Kazemi *et al*. reported a case of an aberrant vein crossing over the ICA bifurcation mimicking an aneurysm on 3DCTA.[8] These reported cases suggest that bleeding sources could lead physicians astray from actual

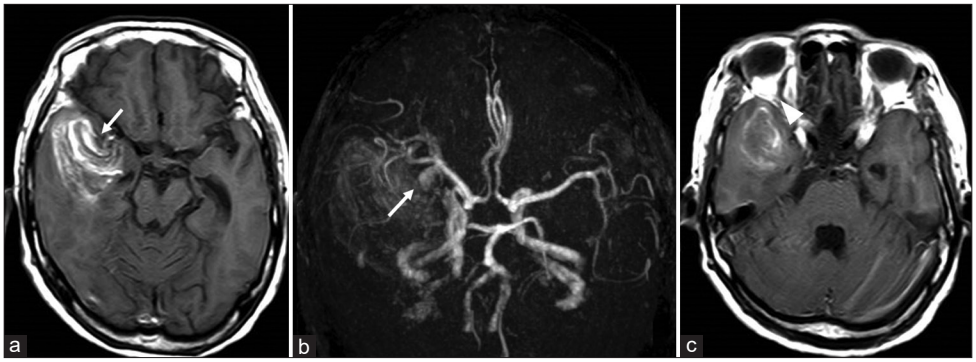

Figure 2: (a) T1-weighted magnetic resonance imaging showing intracerebral hemorrhage and a rounded mass in contact with the right middle cerebral artery (arrow). (b) MR angiography showing an aneurysmal rounded mass arising from the middle cerebral artery bifurcation (arrow). (c) T1-weighted magnetic resonance imaging showing a dilated right superior orbital vein (arrowhead).

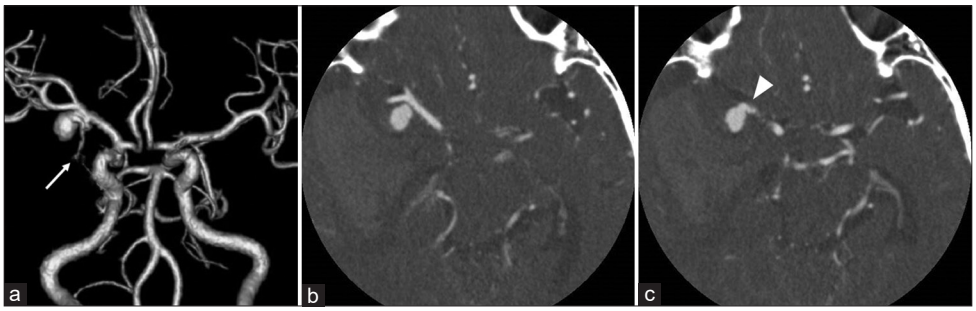

Figure 3: (a) Three-dimensional computed tomography angiography (3DCTA) showing an aneurysmal rounded mass in contact with the middle cerebral artery bifurcation. (b and c) The aneurysmal rounded mass was separated from the middle cerebral artery on axial source image of CTA and a tortuous vessel arising from the aneurysmal mass (arrow).

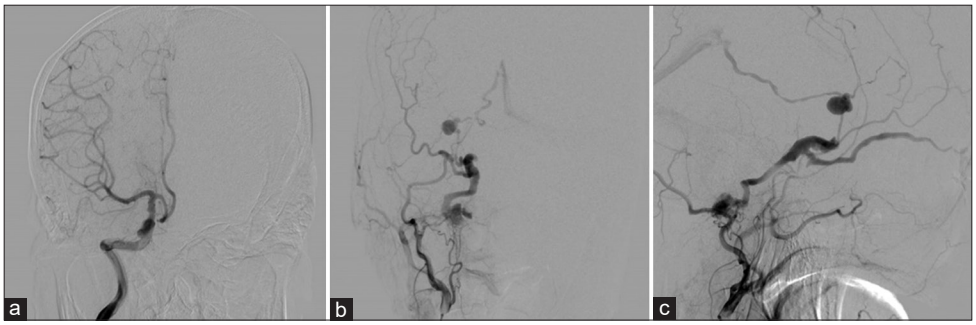

Figure 4: (a) A right internal carotid angiogram, anteroposterior view, showing no aneurysm. A right external carotid angiogram showing a transverse-sigmoid sinus arteriovenous fistula with the varix, supplied by the occipital artery drained into the straight sinus and right superior ophthalmic vein, (b) anteroposterior view, and (c) lateral view.

etiologies, especially if their shapes and locations are similar to other common diseases.

DAVF sometimes has a retrograde cortical venous drainage with varix and this condition is well known for its high risk of hemorrhagic stroke.[2] To the best of our knowledge, only five cases of varices associated with DAVF mimicking cerebral aneurysm have been reported [Table 1], all with variable locations: three were near the anterior communicating artery, one was near the posterior inferior cerebellar artery, and one was near the MCA.[1,3,11,13] Initial diagnostic imaging, such as MRA and 3DCTA, suggested an aneurysm in all cases. DSA was effective in most cases to reach the final diagnosis but it was difficult to make a diagnosis even with DSA in one case.[1] Kwon *et al*. reported that MRA source images may show a flow void cluster corresponding to a DAVF that might be helpful for DAVF identification.[10]

Table 1: Cases of varices mimicking cerebral aneurysms.

Author and year	Age/sex	Symptom	Type of stroke	Location of varix	Final diagnosis	Initial treatment	Retreatment
Machida *et al.*, 1993[11]	48/M	Headache, loss of consciousness	ICH, SAH	Near PICA	TS DAVF	Craniotomy, hematoma removal	TAE
Ogawa *et al.*, 1996[13]	57/M	Visual acuity, homonymous hemianopia	ICH	Near AcomA	Ethmoidal DAVF	Craniotomy, fistula disconnection	No
Chen *et al.*, 2010[1]	41/M	Headache, vomiting	ICH, IVH	Near AcomA	Ethmoidal DAVF	Craniotomy, fistula disconnection	No
Cohen *et al.*, 2015[3]	18/F	Headache, nausea	SAH	Near AcomA	Ethmoidal DAVF	TAE	No
Our case	65/F	Headache, hemiplegia	ICH	Near MCA bifurcation	TS DAVF	Craniotomy, fistula disconnection	No

AcomA: Anterior communicating artery, DAVF: Dural arteriovenous fistula, F: Female, M: Male, ICH: Intracerebral hemorrhage, IVH: Intraventricular hemorrhage, PICA: Posterior inferior cerebellar artery, SAH: Subarachnoid hemorrhage, TAE: Transarterial embolization, TS: Transverse-sigmoid sinus

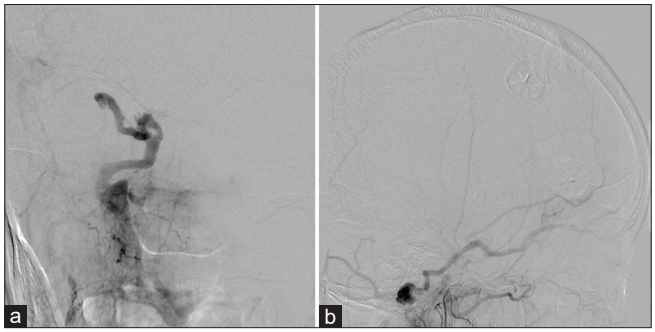

Figure 5: Postoperative digital subtraction angiography showing the residual fistula regurgitated into the right superior orbital vein, (a) anteroposterior view and (b) lateral view.

It is crucial to keep in mind the possibility of various causes for patients with hemorrhagic stroke and carefully interpret data from multiple modalities, including source images, while neurological examination is required for correct pathophysiological diagnosis.

CONCLUSION

This instructive case reminds clinicians to keep in mind that a detailed neurological and radiological examination is essential in obtaining an accurate diagnosis, especially if the bleeding source is similar in shape and location to common lesions (such as a cerebral aneurysm).

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Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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Conflicts of interest

There are no conflicts of interest.

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