DIAGNOSTIC CHALLENGE

The Case of the Man with Blood on the Brain

Joël Surette,[1] BSc, MD 2003 and Gerald Schaller,[2] MD
[1]Faculty of Medicine, Dalhousie University, Halifax, N.S.
[2]Department of Radiology, QEII Health Sciences Centre, Halifax, N.S.

A 20-year-old man presents to the emergency department one afternoon with a severe headache that he describes as the "worst of [his] life." The headache began yesterday while he was doing his laundry. The onset was sudden, the pain severe and associated with nausea and vomiting. He had some mild neck tenderness but no photophobia or diplopia, no weakness, and no numbness. He went to bed because the headache was so bad, however, it became worse this morning prompting him to come to the emergency department. Past medical history is significant for meningitis at age 6. He is on no medications and reports no allergies. Exam shows a sleepy but easily roused young man. Pupils are equal and reactive to light. There is full extraocular motion, normal visual fields, normal CN V-XII, a supple neck and mild left pronator drift. Grip strength is 5/5 bilaterally in upper and lower extremities. Sensation is grossly normal to light touch. Reflexes are equal bilaterally. Cerebellar exam is normal. The lumbar puncture is positive for blood; CT shows an intracranial hemorrhage (Figure 1).

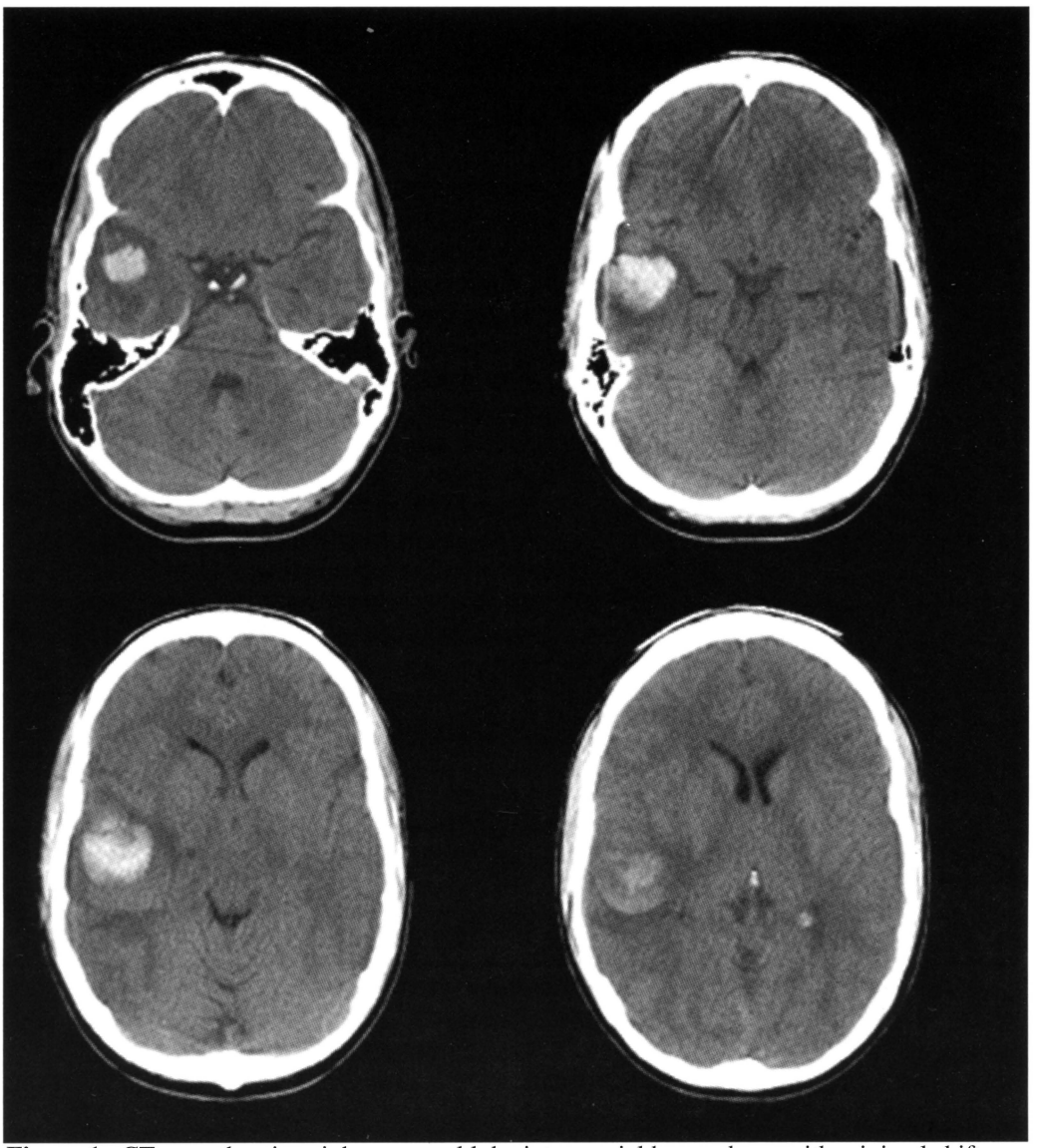

Figure 1. CT scan showing right temporal lobe intracranial hemorrhage with minimal shift and some edema.

Q1: What is the differential diagnosis of intracranial bleeding?

Q2: What is the likely diagnosis in this case?

Q3: What further investigations would you like to order?

Q4: What are the treatment options?

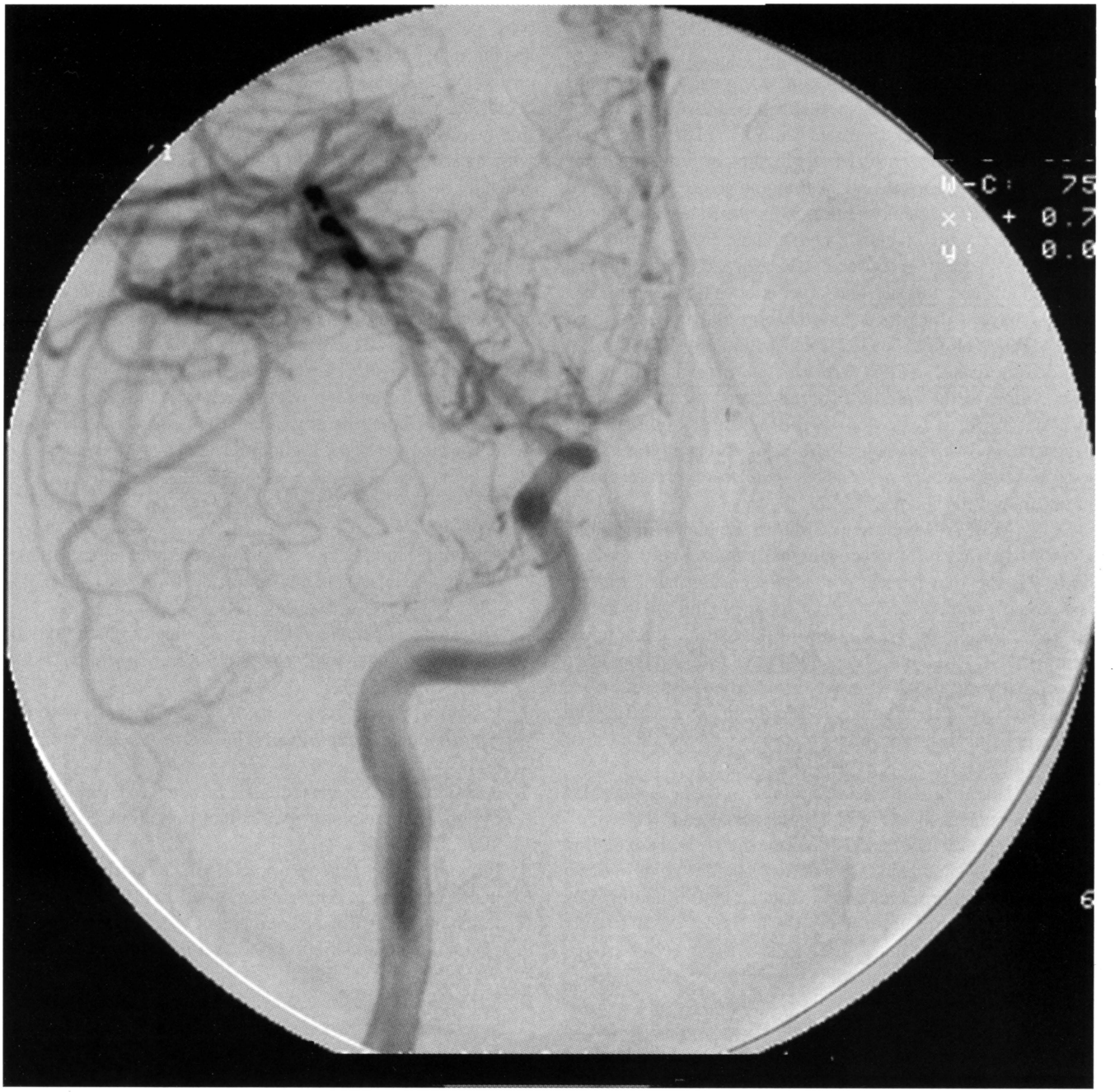

Figure 2. Angiogram of the right internal carotid artery and branches showing a nidus of vessels within the right temporal lobe which appear to be supplied by at least two branches of the right middle cerebral artery.

ANSWERS

A1: Causes of intracranial hemorrhage include trauma, ruptured aneurysm, arteriovenous malformation (AVM), neoplasm, and hypertensive hemorrhage. The CT scan shown in Figure 1 demonstrates intraparenchymal hemorrhage in the pole of the right temporal lobe. There is likely also some subarachnoid hemorrhage (SAH). Ruptured aneurysm is the cause in 85% of cases of SAH excluding trauma.[1]

A2: Trauma is the most common cause of intracranial bleeding, but presents with a different clinical picture. The "worst headache of my life" description is classic for ruptured aneurysm, but also non-specific-the cause proves relatively innocuous in 90% of patients presenting with this as the only symptom. Because of the patient's young age and absence of a history of trauma or hypertension, an underlying neoplastic lesion is unlikely. Ruptured AVM is the most likely diagnosis.

Arteriovenous malformations (AVMs) are a type of congenital vascular malformation composed of tortuous tangles of arteries connected directly to veins without intervening capillaries (a direct shunt).[2,3] There are three distinct zones within an AVM: the feeding arteries, the nidus ("nest")–the tangle itself, and the draining veins. The involved vessels are histologically abnormal, with thin walls due to poor development of elastic and muscle tissue within the media. There are often secondary changes such as thrombosis, calcification, and fibrosis. The larger arteries feeding the AVM usually have a thickened endothelium and hypertrophied media. When brain tissue is present within an AVM (rare), it is nonfunctional. They are most common within the distribution of the middle cerebral artery. Complications include: rupture followed by intracranial hemorrhage, headache, and various neurologic signs and symptoms from local ischemic damage or "vascular steal" from cerebral cortex in the area of the shunt.

Most AVMs present with intracranial hemorrhage. Hemorrhage may be intraparenchymal, subarachnoid, or often both. Patients may present with headache, nausea, vomiting, and possibly fever (classic signs of subarachnoid hemorrhage) but these symptoms are less severe than with subarachnoid hemorrhage. This may be due to the fact that pressure associated with the AVM is lower than that with a ruptured aneurysm or intracerebral hematoma. Obstructive hydrocephalus may also occur, depending on the location of the lesion.

Intracranial hemorrhage from vascular malformations accounts for 1% of all strokes and 10% of all SAHs. The prevalence of AVMs among the general population is uncertain, but autopsy studies of unselected patients indicate that 4 to 5% harbor some form of vascular malformation; only 10 to 15% of these produce symptoms.[2,4] Small AVMs actually pose a greater threat than larger ones. This is because large AVMs have more severe arterial hypotension and are therefore less likely to hemorrhage.

A3: CT scanning is indicated in all patients for whom intracranial bleeding is suspected, followed by lumbar puncture if CT is negative. CT identifies SAH in up to 95% of cases.[1,5,6] A negative CT does not rule out SAH, therefore lumbar puncture is required with a classic history. In the case of AVMs, more detailed information about the location and course of the involved vessels is required for assessing the lesion and planning treatment. This is most practically accomplished by carotid angiography (see Figure 2). CT and MR angiography are emerging as important tools in the detailed evaluation of AVMs.[7-9]

A4: This patient's headache was treated with morphine and gravol. He is also given phenytoin to reduce the risk of seizure and dexamethasone to reduce the inflammatory response to the subarachnoid blood.

The major therapeutic options for an AVM are: (1) surgical resection, (2) intravascular embolization, and (3) gamma-knife radiosurgery.[10,11] Intravascular embolization involves use of various materials (e.g. coils, glues, plastic spheres) to pack the malformation and obstruct bloodflow to and through it. This option is limited by a high rate of disabling or fatal complications and difficulty obtaining complete occlusion. Gamma-knife radiosurgery uses precisely targeted fine beams of ionizing radiation to destroy abnormal tissue while sparing adjacent normal areas. Resolution of an AVM typically occurs over months to two years following a single treatment. Since this patient's aneurysm has already ruptured and bled, neither of these latter options is available leaving surgery as the definitive management.

The risk of surgical resection depends mainly on the size, location and drainage pattern history of an AVM. Size and vascular architecture determine the risks of embolization therapy and radiotherapy. This patient underwent frontotemporal craniotomy for resection of the temporal lobe AVM. A post-operative angiogram showed no residual nidus; he was discharged in stable condition ten days after admission.

REFERENCES

1. van Gijn, J. and Rinkel, G.J. Subarachnoid haemorrhage: diagnosis, causes and management. *Brain* 2001; 124:249-78.
2. Zabramski, J.M., Henn, J.S., and Coons, S. Pathology of cerebral vascular malformations. *Neurosurg Clin N Am* 1999; 10:395-410.
3. Biousse, V. and Newman, N.J. Intracranial vascular abnormalities. *Ophthalmol Clin North Am* 2001;14:243-64.
4. Anzalone, N., Scomazzoni, F., Strada, L., Patay, Z., and Scotti, G. Intracranial vascular malformations. *Eur Radiol* 1998; 8:685-90.
5. Fontanarosa, P.B. Recognition of subarachnoid hemorrhage. *Ann Emerg Med* 1989; 18:1199-205.
6. Latchaw, R.E., Silva, P., and Falcone, S.F. The role of CT following aneurysmal rupture. *Neuroimaging Clin N Am* 1997; 7:693-708.
7. Summers, P.E., Jarosz, J.M., and Markus, H. MR angiography in cerebrovascular disease. *Clin Radiol* 2001; 56:437-56.
8. Kesava, P.P. and Turski, P.A. MR angiography of vascular malformations. *Neuroimaging Clin N Am* 1998; 8:349-70.

9. Thornton, J., Bashir, Q., Aletich, V.A., Debrun, G.M., Charbel, F., and Mafee, M.F. Role of magnetic resonance imaging and diagnostic and interventional angiography in vascular and neoplastic diseases of the skull base associated with vestibulocochlear symptoms. *Top Magn Reson Imaging* 2000; 11:123-37.
10. Mattle, H.P., Schroth, G., and Seiler, R.W. Dilemmas in the management of patients with arteriovenous malformations. *J Neurol* 2000; 247:917-28.
11. Irie, K. *et al.* Treatment of arteriovenous malformation of the brain—preliminary experience. *J Clin Neurosci* 2000; 7:24-9.