WHAT IS YOUR CALL?

Postpartum thunderclap headache

Two days after giving birth to a healthy baby following an uneventful term pregnancy, a healthy 31-year-old woman experienced a severe, diffuse "thunderclap" headache and generalized seizures. She had not received spinal anesthesia during labour. Her blood pressure was 140/90 mm Hg. Results of routine blood work and urinalysis were normal. A plain computed tomography (CT) scan of the head was normal. Magnetic resonance angiography showed multifocal narrowing of the middle and anterior cerebral arteries bilaterally as well as the basilar artery and posterior cerebral arteries (Figure 1). Lumbar puncture showed clear cerebrospinal fluid with no cells, a protein level of 0.4 g/L and a glucose level of 4 mmol/L. Work-up for systemic vasculitis yielded normal findings.

After 2 weeks, the patient's headaches had completely resolved, and she had no recurrent seizures. Repeat magnetic resonance angiography showed normal intracranial arteries with substantial improvement of the narrowing of the vessels noted previously (Figure 2).

What is your diagnosis?

a. Low-pressure headache
b. Cerebral venous thrombosis
c. Subarachnoid hemorrhage with vasospasm
d. Pre-eclampsia or eclampsia
e. Reversible cerebral vasoconstriction syndrome
f. Vasculitis of the central nervous system

See page 1034 for the diagnosis.

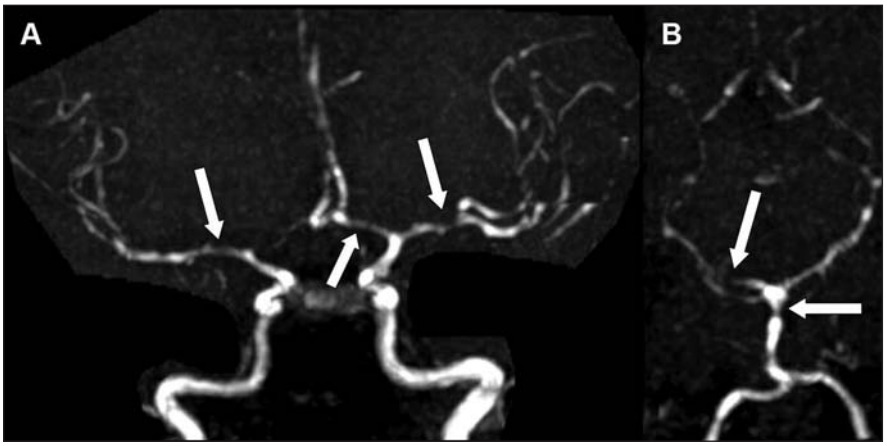

Figure 1: Magnetic resonance angiography of the brain of a woman with thunderclap headache post partum. Multiple areas of narrowing are seen in the middle and anterior cerebral arteries bilaterally (A, arrows) and in the basilar artery and posterior cerebral arteries (B, arrows).

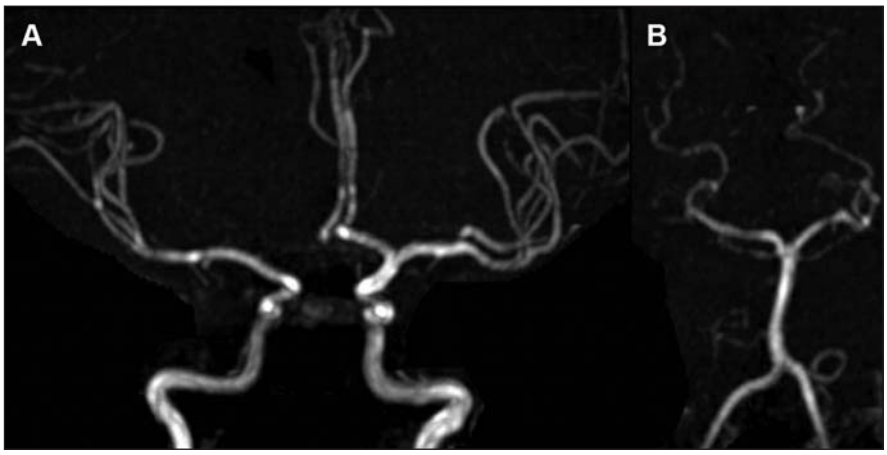

Figure 2: Repeat magnetic resonance angiography performed 2 weeks later showing substantial improvement of the narrowing of the cerebral arteries noted earlier.

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Discussion

On the basis of the patient's clinical history, the results of the lumbar puncture and laboratory tests, and the findings on the serial magnetic resonance angiography, the answer is (e) reversible cerebral vasoconstriction syndrome.

This syndrome includes conditions such as Call–Fleming syndrome and postpartum angiopathy. It is characterized by recurrent thunderclap headache with or without neurologic deficits. The typical patient is a woman between 20 and 50 years of age presenting with thunderclap headache.[1] Men are also affected. Thunderclap headache is recognized as a severe headache reaching peak intensity within 1 minute. In reversible cerebral vasoconstriction syndrome, the headache may be occipital or diffuse. It may be associated with nausea, vomiting and photophobia. However, the headache has features that distinguish it from migraine headache. Generalized seizures may occur during the episode, but epilepsy does not ensue.

Reversible cerebral vasoconstriction syndrome is often idiopathic but may be associated with several conditions, including pregnancy and the puerperium, exposure to certain drugs, and catecholamine-secreting tumours.[1]

The pathophysiology of reversible cerebral vasoconstriction syndrome is unclear but likely includes abnormalities in cerebral vascular tone. The imaging hallmarks are the multifocal areas of constriction and dilatation of cerebral arteries that resolve within days to weeks. The treatment remains empirical and includes observation and possibly calcium-channel blockers.[1] Most importantly, the prognosis for sequelae and recurrence tends to be favourable. However, vasoconstriction, if severe, can cause cerebral infarction, hemorrhage and brain edema, which can lead to poor outcome.

Differential diagnosis: Thunderclap headache is a characteristic symptom of subarachnoid hemorrhage. It can also be the presenting feature of cervical arterial dissection, cerebral venous thrombosis, pituitary apoplexy, low-pressure headache, pre-eclampsia or eclampsia, and reversible cerebral vasoconstriction syndrome (Table 1).

Table 1: Differential diagnosis of postpartum headache

Condition	Clinical features	Associated with thunderclap headache
Tension-type headache	Bilateral, pressure-like headache; no associated symptoms	No
Low-pressure headache	Postural headache, nausea, tinnitus, neck stiffness	Yes
Cerebral venous thrombosis	Focal neurologic deficits, seizures, papilledema	Yes
Subarachnoid hemorrhage	Decreased level of consciousness, nuchal rigidity, subhyaloid hemorrhage	Yes
Pre-eclampsia or eclampsia	Hypertension, proteinuria, seizures	Yes
Cerebral vasculitis	Constant headache, episodes of neurologic deficits	No
Reversible cerebral vasoconstriction syndrome	Confusion, seizures, visual symptoms	Yes

An unenhanced CT of the head should be done to exclude subarachnoid hemorrhage and intraparenchymal hemorrhage. If the CT is negative, lumbar puncture should be performed to rule out "CT-negative" subarachnoid hemorrhage, meningitis and inflammatory diseases. If the results of the lumbar puncture are unremarkable, additional neuroimaging such as magnetic resonance imaging (MRI) and neurovascular imaging should be performed to assess for venous thrombosis, dissection, vasculitis and reversible cerebral vasoconstriction syndrome. Although magnetic resonance angiography and CT angiography of the brain are often used for evaluating the intracranial vessels, catheter-based cerebral angiography remains the "gold standard." However, although cerebral angiography has high sensitivity in diagnosing reversible cerebral vasoconstriction syndrome, it lacks specificity and the clinical picture is critical to establishing the diagnosis.

The incidence of postpartum headache has been reported as being between 11% and 80%.[2] Persistent headache in the postpartum period is most often due to tension-type headache.[3] This headache is bilateral, pressure-like and not thunderclap in onset. Patients with tension-type headache do not have other associated findings. The presence of seizures and the thunderclap headache therefore ruled out tension-type headache in our patient.

The risk of accidental dural puncture while performing epidural insertion during labour is 1 in 67.[4] Low-pressure headache develops in about half of these patients. This type of headache is due to leakage of cerebrospinal fluid. It usually has a postural component, being relieved when the patient is in the recumbent position. Our patient, however, did not receive epidural anesthesia during labour. In patients who receive epidural anesthesia during labour, meningitis must also be considered in the differential diagnosis of headache. Nuchal rigidity, photophobia and vomiting may be present. Lumbar puncture would typically reveal leukocytosis, a high protein level and a low glucose level. The absence of nuchal rigidity and, most importantly, the normal results of lumbar puncture excluded the possibility of meningitis in our case.

Pre-eclampsia must be ruled out in all postpartum patients with headache. These patients present with hypertension and proteinuria in addition to headache. The additional presence of seizures suggests eclampsia. Recognition of this entity is important, since

prompt use of magnesium sulfate prevents recurrence of seizures. Eclampsia was an important consideration in our patient, and we administered magnesium sulfate and phenytoin to prevent seizures. However, the absence of proteinuria and hypertension made the diagnosis of pre-eclampsia or eclampsia unlikely in the present case.

The risk of cerebral venous thrombosis is increased during pregnancy and during the postpartum period because of a relative hypercoagulable state. The headache is usually severe and can be of the thunderclap type. Vomiting, focal neurologic deficits and seizures may be accompanying features. Magnetic resonance imaging readily demonstrates occlusion of the dural sinus and possible venous infarction. Venography reveals occlusion of the dural sinus or cortical veins.

Subarachnoid hemorrhage due to rupture of an intracranial aneurysm classically presents with thunderclap headache and must be ruled out. Accompanying symptoms may be vomiting, decreased level of consciousness and focal neurologic deficits. On examination, patients usually have nuchal rigidity. Normal findings on the plain CT of the head and findings from the lumbar puncture and subsequent magnetic resonance angiography ruled out cerebral venous thrombosis and subarachnoid hemorrhage in our patient.

Vasculitis of the central nervous system can cause multifocal narrowing of intracranial arteries. Hence, patients such as ours should be extensively investigated for vasculitis. Vasculitis of the central nervous system usually presents with a progressive, indolent headache but not of the thunderclap variety. Episodes of neurologic deficits often occur. Results of lumbar puncture are frequently abnormal, unlike with reversible cerebral vasoconstriction syndrome. Work-up for vasculitis includes laboratory tests such as erythrocyte sedimentation rate, measurement of the C-reactive protein level, tests for antinuclear antibodies and antineutrophil cytoplasmic antibodies, and cerebral angiography. Magnetic resonance imaging may show multiple infarcts of varying ages. Angiography shows areas of stenoses or occlusions involving single or multiple arteries. The abnormalities are irreversible. The presence of thunderclap headache, normal laboratory results for vasculitis and, most importantly, reversibility of vascular narrowing, ruled out vasculitis of the central nervous system in our case.

As is often the case, reversible cerebral vasoconstriction syndrome was diagnosed only in retrospect in our patient. The initial clinical presentation with thunderclap headache and seizures suggested eclampsia. However, there was no proteinuria or significant hypertension. Magnesium sulfate and phenytoin were given to prevent seizures, although the recommended treatment for eclampsia is magnesium sulfate alone. Another possibility we considered was subarachnoid hemorrhage, but the noncontrast CT of the head and the results of the lumbar puncture ruled it out. The presence of multiple areas of narrowing of the intracranial arteries on the magnetic resonance angiography suggested vasculitis. Even with a normal vasculitis panel, vasculitis of the central nervous system cannot be completely excluded. However, the patient presented with thunderclap headache, which is unusual for cerebral vasculitis. Rapid resolution of clinical symptoms and narrowing of the intracranial vessels helped us confirm the final diagnosis.

The diagnosis of reversible cerebral vasoconstriction syndrome should be entertained in patients with thunderclap headache in the presence of narrowing of intracranial vessels, especially when other causes have been ruled out, the diagnosis is often retrospective and can be confirmed only after documented resolution of clinical symptoms and vascular narrowing.

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REFERENCES

1. Calabrese LH, Dodick DW, Schwedt TJ, et al. Narrative review: reversible cerebral vasoconstriction syndromes. *Ann Intern Med* 2007;146:34-44.
2. Scharff L, Marcus DA, Turk DS. Headache during pregnancy in the post-partum: a prospective study. *Headache* 1997;37:203-10.
3. Stella CL, Jodicke CD, How HY, et al. Post-partum headache: Is your work-up complete? *Am J Obstet Gynecol* 2007;196:318.e1-7.
4. Turnbull DK, Shepherd DB. Post-dural puncture headache: pathogenesis, prevention and treatment. *Br J Anaesth* 2003;91:718-29.

CMAJ invites submissions to "What is your call?" Clinical details (including images) are presented on the first page along with a multiple-choice question about the diagnosis. The answer and a brief discussion of the condition follow on the second page. We specifically invite submissions illustrating common or important radiographic and electrocardiographic diagnoses of appeal to a general audience. We allow up to 5 references and require authors to obtain consent from the patient for publication of his or her story (form available at www.cmaj.ca/authors/checklist.shtml). Submit manuscripts online at http://mc.manuscriptcentral.com/cmaj.