Neurology Case Studies Cerebrovascular Disease

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KEYWORDS

- Intracranial stenosis Intracranial occlusive disease Migrainous stroke
- Susac syndrome Cerebral autosomal dominant arteriopathy with subcortical
- infarcts and leukoencephalopathy Posterior reversible encephalopathy syndrome
- Eclampsia

KEY POINTS

- This article provides a series of interesting neurologic cases practice and discusses diagnosis and treatment.
- Because several of the cases are uncommon causes of stroke, there is no high-level evidence base to guide the clinician in relation to diagnosis and treatment.
- One must recognize the pattern of clinical features to arrive at the proper diagnostic workup and diagnosis and exercise good clinical judgment in relation to reasonable treatment.
- Careful neurologic history and examination remain paramount to determining the correct diagnosis and subsequent treatment.

CASE 1: MANAGEMENT OF SYMPTOMATIC INTRACRANIAL ARTERY STENOSIS Case Presentation

A 79-year-old man with a medical history of hypertension, type-2 diabetes mellitus, hyperlipidemia, and coronary artery disease presented with spells of intermittent left hand grip weakness and left hand numbness. His wife observed facial asymmetry and speech impairment with some of the episodes. The duration of the spells was up to 1 hour, and they occurred over a 2- to 3-week period. In the emergency department (ED), the National Institutes of Health Stroke Scale (NIHSS) score was 1 (left upper extremity sensory impairment). The patient did not receive intravenous (IV) thrombolytic therapy because of delay in presentation to the ED. He had no history of cardiac

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arrhythmia such as atrial fibrillation. His low-density lipoprotein cholesterol level was 111 mg/dL, and HbA1c was 7.4%. The patient had computerized axial tomography (CT) and MRI of the brain. Brain CT was unremarkable, but brain MRI showed areas of restricted diffusion (Fig. 1A, B). The areas of restricted diffusion were interpreted as being consistent with embolic stroke. There was no evidence of hemodynamically significant stenosis of the carotid arteries. Magnetic resonance angiography (MRA) of the brain without contrast was limited by motion artifact. CT angiography (CTA) was not obtained, as there was renal impairment caused by diabetes. Echocardiography with bubble study did not show evidence of a cardiac source of stroke. The patient was discharged to home on aspirin, 81 mg daily, and atorvastatin, 40 mg at bed time. He underwent cardiac event monitoring for 28 days, and there was no evidence of atrial fibrillation.

The patient was readmitted to the hospital almost 4 weeks later with left arm and leg weakness, decrease sensation of the left arm, dysarthria, and left facial droop. His symptoms improved, and in the ED his NIHSS score was 3 (left arm and leg drift and left arm sensory loss). In spite of previous stroke education, there was a delay in the patient's presentation to the ED, and he did not receive IV tissue plasminogen activator therapy. Brain CT in the ED did not show an acute intracranial process. Brain MRI again showed multiple new areas of restricted diffusion consistent with embolic strokes (Fig. 1C, D). The patient had conventional cerebral angiography, which showed hemodynamically significant stenosis of the right M1 segment of the middle cerebral artery (MCA) (Fig. 2A, B).

Despite intensive medical management with clopidogrel, 75 mg/d, aspirin, 81 mg/d, and atorvastatin, 80 mg/d, the patient had fluctuations of neurologic symptoms and signs that were blood pressure (BP) dependent and required IV fluids and vasopressors for 2 days. His examination findings remained stable after 2 days, and he was discharged to an acute rehabilitation hospital with an intensive risk factor control regimen. The BP target for this patient was 130/80 mm Hg.

The patient had a third hospital admission for worsening neurologic symptoms about 4 weeks later. He had pronounced left-sided weakness. His NIHSS score was 8. Brain CT did not show an acute process. Brain MRI showed multiple new areas of restricted diffusion (Fig. 1E, F). With a relative decrease in BP, the patient had significant fluctuation and worsening of focal neurologic deficits during hospitalization. His BP ranged from 160/84 mm Hg to 126/68 mm Hg.

Clinical Questions

- 1. What is the diagnosis in this patient?
- 2. What is the significance of BP variability and BP augmentation in this patient?
- 3. According to current American Heart Association/American Stroke association guidelines, how should this patient be treated?

Discussion

Recurrent cerebral ischemic symptoms in a single vascular territory, such as in this patient, suggest a high-grade focal large artery stenosis. The symptoms can be caused by cerebral hypoperfusion or artery-to-artery embolism. In this patient, there was no evidence of a high-risk source of cardiac embolism. Given the constellation of symptoms and neuroradiologic findings, we believe that intracranial occlusive disease with artery-to-artery embolism was the most likely cause of stroke.

Intracranial occlusive disease is common in many Asian countries where transcranial Doppler may be used for detection of microembolic signals.^{1–5} Therefore, noninvasive imaging by means of CTA, MRA, or transcranial Doppler of the intracranial vasculature is recommended to exclude the presence of proximal intracranial stenosis or occlusion (class I; level of evidence A).⁶

Vascular Neurology Case Studies

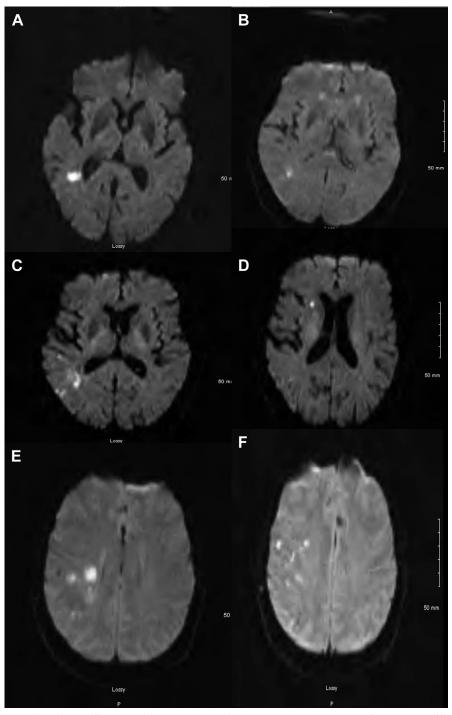


Fig. 1. (A-F) MRI of the brain (axial sequence) DWIs show multiple areas of restricted diffusion in the brain in the right MCA territory at different time points according to the patient's symptom course over an approximately 8-week period.

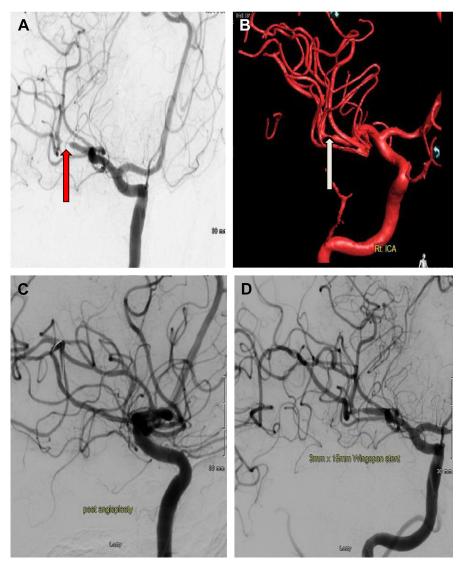


Fig. 2. (*A* and *B*) Conventional cerebral angiography shows significant stenosis of distal right MCA M1 segment. (*C*) Conventional cerebral angiography shows improvement in right MCA stenosis after angioplasty. (*D*) Conventional cerebral angiography shows patency of right MCA after Wingspan stent placement. An *arrow* delineates the key neuroradiological feature referred to in the respective figure segment.

Blood pressure variability in acute stroke patients with intracranial atherosclerotic occlusive disease may be associated with neurologic deterioration and poor neurologic outcome.⁷ Therefore, augmentation of BP and reduction of BP fluctuations in the acute setting may be useful, especially when auto-regulatory mechanisms are compromised in the area of cerebral infarction and the surrounding penumbra. The penumbra may be thought of as a shadow zone of compromised cerebral blood flow or perfusion in which brain tissue is rendered ischemic and dysfunctions, but frank infarction has not occurred. The rationale to augment BP is to salvage the ischemic penumbra. Elevation of BP may augment collateral blood flow.^{8,9}

Our patient was treated with intravenous fluids and required vasopressors to elevate the BP and to avoid further fluctuations in neurologic status. Systolic BP was then maintained at greater than 150 mm Hg, and worsening of neurologic status ceased. However, with attempted weaning of vasopressors, there was recurrence of neurologic signs and symptoms.

For patients with recent stroke or transient ischemic attack (ie, within 30 days) attributable to severe stenosis (70%–99%) of a major intracranial artery, the addition of clopidogrel, 75 mg/d, to aspirin therapy for 90 days may be reasonable (class IIb; level of evidence B).¹⁰ Our patient was on dual antiplatelet and statin therapy at the time of his third admission to the hospital. Aggressive medical management is found to be efficacious when there is high-grade (70%–99%) symptomatic intracranial large artery occlusive disease according to findings from the SAMMPRIS trial.¹¹ However, the Wingspan stent system is not recommended as an initial treatment (class III; level of evidence B).¹⁰ Because of hemodynamic compromise, our patient underwent angioplasty and stenting of the right MCA stenosis. The procedure was successful, and there were no complications (**Fig. 2C**, D). Thus, although angioplasty and stenting of major intracranial arteries is not generally recommended in practice, when there is a high-grade stenosis and inability to wean a patient from vasopressors because of hemodynamic compromise, and stenting may be indicated.

CASE 2: AN INTERESTING CASE OF MIGRAINE HEADACHE AND STROKE Case Presentation

A 27-year-old woman presented to the ED with left temporal throbbing headache, left facial and left arm numbness, and left arm weakness. She had intractable headaches with some fluctuation for the past 3 to 4 weeks. The focal neurologic symptoms lasted for 30 minutes and then completely resolved. Her BP was 120/70 mm Hg. She had a history of migraine headache but never had focal neurologic symptoms. The patient was not pregnant and had never taken birth control pills. There was no history of cigarette smoking. Brain MRI did not show an acute intracranial process. The diffusion-weighted images (DWI) were unremarkable. In the ED the patient was treated with sumatriptan injection, and dihydroergotamine mesylate (DHE) was administered during acute hospitalization over a 12-hour period. Her symptoms completely resolved within 24 hours of hospitalization.

However, the next day she woke up and could not see off to the left side. There was a left visual field defect but no other focal neurologic impairment. The patient had a repeat brain MRI and MRA. MRI brain showed findings consistent with right occipital ischemic stroke, and brain MRA showed narrowing of the right posterior cerebral artery (PCA) (Fig. 3A, B). Diagnostic studies for blood markers of inflammation, hyper-coagulable testing, and cerebrospinal fluid (CSF) analysis were unremarkable. Her BP was not elevated at any time during hospitalization or stay in the ED. She was head-ache free at the time of discharge but had a visual field defect. Repeat brain MRA after 3 months was unremarkable, and there were no findings suggestive of vasculitis. A clinically significant visual field deficit was still present 1 year later.

Clinical Questions

- 1. What is the diagnosis in this case?
- 2. What is the mechanism of stroke?
- 3. Is it safe to administer sumatriptan and DHE concurrently in a migraine patient to abort an acute headache?

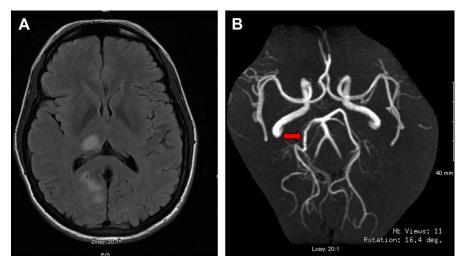


Fig. 3. (*A*) MRI of the brain (axial sequence) FLAIR image shows multiple hyperintensities in the right PCA distribution involving occipital lobe and thalamus. (*B*) MRA of the brain shows irregularity of right PCA concerning for possible vasculitis or other vasculopathy. An *arrow* delineates the key neuroradiological feature referred to in the respective figure segment.

Discussion

The differential diagnosis in this case includes migrainous infarction, vasculitis, hypercoagulable condition, posterior reversible encephalopathy syndrome (PRES), and a complication related to medications administered during hospitalization to treat the headache. The diagnostic workup, including laboratory studies and CSF analysis, was not suggestive of vasculitis or a hypercoagulable condition. Moreover, repeat brain MRA after 3 months was essentially unremarkable. The patient declined to have a conventional cerebral angiography study. PRES was unlikely, as BP was normal throughout the hospital stay. Therefore, it is thought that the patient may have had stroke related to sumatriptan and DHE, used to treat the headache, although the mechanisms underlying cerebral ischemia associated with migraine are many (eg, vasospasm, endothelial injury, platelet aggregation, prothrombotic state, cortical spreading depression, cerebral arterial dissection, genetic variants, and traditional vascular risk factors).¹²

One should be cautious when using vasoconstrictive medications in patients with migraine cephalalgia. Triptans may be safe if used to treat migraine even with associated neurologic symptoms.^{13,14} However, these medications should be used with caution, especially in combination with other drugs that can cause vasoconstriction such as DHE. This may deserve additional emphasis if the patient has an underlying disorder predisposing to stroke such as vasculitis or hypercoagulable condition. DHE is an ergot alkaloid and has potential for vasoconstriction; therefore, it may be preferred not to use multiple medications concurrently (eg, at least within 24 hours) that have such properties.¹⁵ However, there are no clear guidelines to support the latter timeframe.

CASE 3: A PATIENT WITH RETINAL ARTERY OCCLUSIONS AND OTITIC NERVOUS SYSTEM INVOLVEMENT Case Presentation

A 34-year-old woman presented with visual disturbance, headaches, intermittent vertiginous feeling, and behavioral changes for almost 2 years. She had progressive

worsening of visual acuity and blurry vision but never had complete or sudden visual loss. The patient described a vertiginous feeling as being lightheaded or as a spinning sensation.

She was initially diagnosed with amaurosis fugax at an outside hospital because of visual impairment. However, she never had sudden monocular visual loss but rather had progressive visual impairment over time. She was treated with aspirin, 81 mg/d, and had stroke diagnostic studies before being examined in our clinic. The diagnostic studies including those of the cervical and intracranial vasculature were unremarkable. Transesophageal echocardiography found a patent foramen ovale. Her patent foramen ovale was closed at an outside hospital at the recommendation of other consultants.

Her family reported a few episodes of acute confusion in the last year. They also reported that her interpersonal interactions were abnormal. The patient stopped working and driving because of her episodic confusion, behavior change, and difficulty with balance and hearing.

Brain MRI was done at an outside hospital and showed multiple hyperintensities on T2 fluid-attenuated inversion recovery (FLAIR) images involving the corpus callosum (Fig. 4). The patient was referred for an ophthalmologic evaluation, which found bilateral and multiple branch retinal artery occlusions on fluorescein angiography. Later she had an otolaryngology evaluation, which confirmed bilateral sensorineural hearing loss.

Clinical Questions

- 1. What is the diagnosis in this case?
- 2. What is the cause of visual symptoms in this patient, and why was a careful neurologic history so important in this case?
- 3. Was there any indication for oral anticoagulation therapy in this patient?

Discussion

The differential diagnosis in this case includes a demyelinating disorder such as multiple sclerosis, vasculitis, ischemic stroke, and an endotheliopathy such as Susac syndrome. Lesions predominantly of the corpus callosum are not common and are not pathognomonic of multiple sclerosis, vasculitis, or stroke. Based on the neuroradiologic findings,

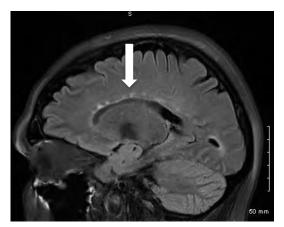


Fig. 4. MRI of the brain (sagittal sequence) T2 (FLAIR) image shows multiple hyperintensities involving the central fibers of the corpus callosum and typical of a neuroradiologic diagnosis of Susac syndrome. An *arrow* delineates the key neuroradiological feature referred to in the respective figure segment.

one should consider a diagnosis of Susac syndrome, as lesions involve the central fibers of the corpus callosum, sparing the more peripheral ones.¹⁶ Patients with Susac syndrome have associated visual and hearing impairment as seen in our patient.

Susac syndrome is an immune-mediated endotheliopathy. It is characterized by the triad of encephalopathy, cochlear hearing loss, and retinal artery occlusions. The clinical course of Susac syndrome is usually self-limited and fluctuating and lasts from 2 to 4 years, but duration varies. Some patients recover with little or no residual issues, but others can be profoundly impaired. Options for therapy include corticosteroids, intravenous immunoglobulin, and other immunosuppressant drugs.^{17,18}

This patient had visual symptoms caused by the involvement of retinal arterial occlusions and not by amaurosis fugax. A careful neurologic history is important when making a diagnosis of amaurosis fugax. Patients with amaurosis fugax usually have sudden loss of vision in one eye that may be described as a shade coming over the eye or from the side and as a blackening, greying, or clouding of vision. Visual loss is not progressive over weeks or months but rather typically occurs over seconds to minutes or longer.

In this patient, the clinicians stopped antiplatelet therapy and administered oral anticoagulation. Use of oral anticoagulation is generally reserved for patients with certain types of cardiac source embolism (eg, atrial fibrillation), certain hypercoagulable states (eg, antiphospholipid antibody syndrome), and deep vein thrombosis or pulmonary embolism who have stroke.¹⁰

CASE 4: INTRACRANIAL OCCLUSIVE DISEASE Case Presentation

A 49-year-old woman with a medical history of bipolar disorder had an episode of lightheadedness, dizziness, visual disturbance, confusion, and impaired level of consciousness during a trip to Paris. The episode lasted for 10 to 15 minutes with complete resolution of symptoms except for a mild headache, which lasted for 1 to 2 hours. The patient had no medical history of headache or similar symptoms in the past. She underwent brain MRI and MRA of the head and neck in Paris and was informed that all of the imaging findings were unremarkable. She underwent hypercoagulable workup, echocardiography, and electroencephalography on her return to the United States and was told that all diagnostic findings were unremarkable.

She had 2 more episodes of similar symptoms within 1 month of her return to the United States. The spells lasted longer than her first episode. She also had associated difficulty with speech and mild left temporal headache. The patient had vascular migraine headache diagnosed and was advised to take topiramate. Topiramate was administered for several months at a dose of 100 mg twice a day, but the patient continued to have 1 to 2 episodes of focal symptoms every month. She was referred to our vascular neurology clinic for a second opinion. Brain MRI findings were normal, but MRA of the head and neck carried out in Paris showed significant vascular abnormalities. She underwent CTA of the head and neck and conventional cerebral angiography. These studies showed complete occlusion of the internal carotid arteries on both sides and a vascular network of collateral circulation at the base of the brain (Fig. 5).

Clinical Questions

- What is the diagnosis in this case?
- 2. Does this disease entity occur worldwide and in adults preferentially?
- 3. What are the surgical options for these patients?

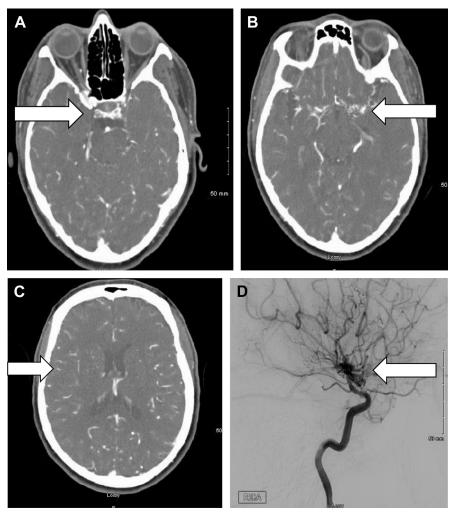


Fig. 5. (*A*) CTA of the brain shows complete occlusion of both internal carotid arteries. (*B*) CTA of the brain shows abnormal vascular network at the base of the brain. (*C*) CTA of the brain shows dilated collateral blood vessels close to the brain surface that are normally not seen in healthy individuals. (*D*) Conventional cerebral angiography of the brain shows abnormal vascular network known as *puff of smoke* (moyamoya). An *arrow* delineates the key neuroradiological feature referred to in the respective figure segment.

Discussion

The differential diagnosis in this case includes cerebral ischemia, vascular migraine, seizure disorder, autonomic instability, and cardiac arrhythmia. In retrospect, the initial neuroimages from France were not normal, emphasizing the potential importance of personally reviewing neuroimages or reviewing them with a neuroradiologist when possible. Based on the neurologic and neuroradiologic findings, a diagnosis of Moyamoya syndrome was made. If a patient with vascular migraine continues to have focal symptoms in spite of taking prophylactic medication, consider ordering imaging of the cervical and intracranial vasculature to rule out vascular pathology.

There is some misconception about the prevalence of Moyamoya disease. Although Moyamoya syndrome is common in some persons of Asian heritage, it can be seen in whites.¹⁹ It is a chronic, occlusive cerebrovascular disease of unknown etiology. It is characterized by bilateral steno-occlusive changes at the terminal portion of the internal carotid arteries. These patients also develop an abnormal vascular network at the base of the brain with extensive collaterals to compensate for main vessel occlusion. The collateral circulation at the base of the brain may look like a puff of smoke or what is referred to as *moyamoya* in Japanese. The disease primarily affects children, but it can also occur in adults. Children may present with cerebral ischemic symptoms after crying, as hyperventilation may cause cerebral artery constriction and trigger a reduction of cerebral blood flow given tenuous collateral blood flow.

Moyamoya syndrome can lead to ischemic or hemorrhagic strokes. Surgical revascularization procedures may help prevent ischemic vascular complications in these patients. These procedures include direct revascularization—superficial temporal artery middle cerebral artery bypass—and indirect revascularization—encephaloduro-arterio-synangiosis and encephalo-myo-synangiosis. Our patient underwent encephalo-duro-arterio-synangiosis on the right side and had the same procedure on the left side after 3 months. She tolerated these procedures well and is symptom free for the last 1 year and has been free of disability.²⁰

CASE 5: AN IMPORTANT HERITABLE CAUSE OF STROKE AND VASCULAR DEMENTIA Case Presentation

A 41-year-old woman with a history of traumatic brain injury, tinnitus, chronic daily headaches, and seizure disorder was evaluated for worsening memory, mood disorder, unsteady gait, and balance problems. She had no history of vascular risk factors such as hypertension, hyperlipidemia, or diabetes mellitus. MRI of the brain showed multiple hyperintensities on T2 FLAIR images with involvement of the tips of temporal lobes (Fig. 6).

She underwent extensive diagnostic workup for markers of inflammation and demyelination. Study findings were normal, including CSF analysis, with the exception of syphilis antibody titer, which was elevated, and Treponema pallidum antibodies, which were reactive. Her CTA scans of the head and neck were unremarkable. She was evaluated by an infectious disease consultant, and meningovascular syphilis was diagnosed, which was treated with intravenous penicillin therapy. The patient did not have improvement in her symptoms over time and continued to have headaches, memory and mood disorder, and intermittent focal neurologic symptoms. Based on MRI brain findings, especially involvement of the tips of the temporal lobes, and clinical features, genetic testing was obtained. The results showed the presence of a NOTCH 3 gene mutation on chromosome 19.

Clinical Questions

- 1. What is the diagnosis in this case?
- 2. Is there curative or high-level evidence-based preventive therapy for this disorder?
- 3. Can these patients be treated with thrombolytic therapy if they have an acute ischemic stroke?

Discussion

The differential diagnosis in this case includes stroke, vasculitis, a demyelinating disorder such as multiple sclerosis, neurosyphilis, and cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL). One should consider a diagnosis of CADASIL when there is a history of migraine headache, absent

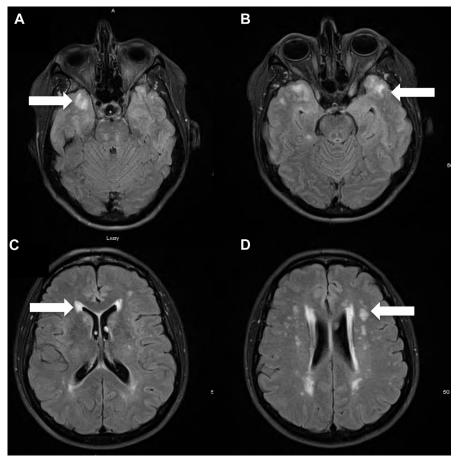
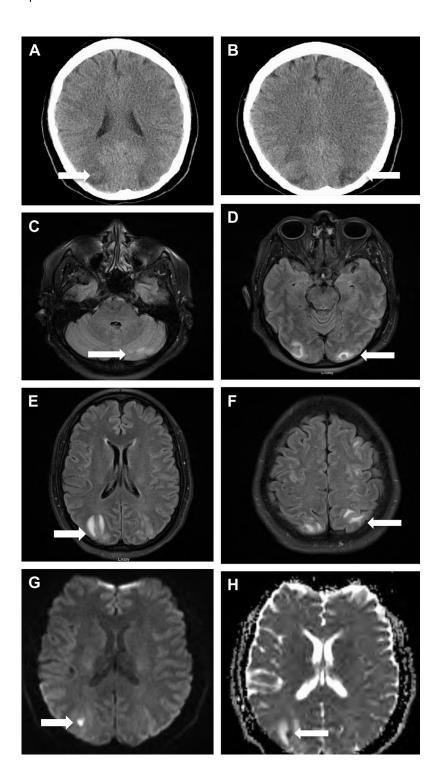


Fig. 6. (*A*, *B*) MRI of the brain (axial sequence) T2 (FLAIR) image shows multiple hyperintensities involving the white matter of the tips of temporal lobes. (*C*) MRI of the brain (axial sequence) T2 (FLAIR) image shows multiple hyperintensities involving the periventricular white matter. (*D*) MRI of the brain (axial sequence) T2 (FLAIR) image shows multiple hyperintensities involving the subcortical white matter. An *arrow* delineates the key neuroradiological feature referred to in the respective figure segment.

or few vascular risk factors (eg, cigarette smoking may be present), mood disorder, and neurologic symptoms and signs. Characteristically, there is involvement of the white matter of the tips of temporal lobes on brain MRI. This patient underwent genetic testing and had a NOTCH 3 gene mutation confirming the diagnosis of CADASIL. This mutation is expressed predominantly in vascular smooth muscle. There is progressive degeneration of the smooth muscle cells in blood vessels, and the vasculopathy is generalized. Mutations in the NOTCH 3 gene cause abnormal accumulation of protein in vascular smooth muscle cells in the cerebral and extracerebral vessels including the skin. On microscopic examination there are osmophilic granules in the blood vessel wall.

Clinically, CADASIL is characterized by history of migraine headache, recurrent ischemic strokes, cognitive decline, and psychiatric affective symptoms. CADASIL begins with migraine with aura in almost one-third of patients. It is the most common heritable cause of stroke and vascular dementia in adults. These patients have transient

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ischemic attacks and strokes between the ages of 30 and 50 years. There are multiple, small, deep cerebral infarcts, and neuroimaging findings are consistent with leukoencephalopathy. During the time course of CADASIL, women earlier on may have fewer small deep infarcts compared with men. Frank dementia is usually a late complication. Diagnostic studies include MRI of the brain and skin biopsy or genetic testing. The NOTCH 3 gene mutation test is the favored means to establish a diagnosis.^{21–24}

There is no definitive prevention or treatment for CADASIL. However, antiplatelet drugs rather than anticoagulants are often administered. The role of calcium channelblocking agents and other BP-lowering medications with vessel wall-protective properties are often used though remain to be formally tested in a large-scale clinical trial.²³

If a CADSIL patient has an acute ischemic stroke, thrombolytic therapy may be administered. However, safety of tissue plasminogen activator in patients with CADASIL is not well characterized, and there is no definite recommendation about CADASIL patients and administration of acute thrombolytic therapy for ischemic stroke in current American Heart Association/American Stroke guidelines.

CASE 6: AN INTERESTING CLINICO-NEURORADIOLOGIC ENTITY IN PREGNANCY Case Presentation

A 35-year-old woman, gravida 2, para 2, aborta 0, 5 days postpartum after an uncomplicated vaginal delivery presented to the ED after multiple seizure spells. She had 2 witnessed tonic-clonic seizure spells at home and one more spell en route to the hospital and a prolonged postictal state. The patient had a mild continuous retro-orbital headache since delivery and had blurry vision with worsening of headache. Her BP in the ED was 187/119 mm Hg. She was started on IV nicardipine and magnesium sulfate.

CT of the brain showed areas of hypodensities involving the occipital lobes (Fig. 7A, B), and brain MRI showed multiple hyperintensities on T2 FLAIR images, which were mainly located posteriorly (Fig. 7C–F). The DWI images showed areas of hyperintensities and corresponding areas on apparent diffusion coefficient study consistent with ischemic injury (Fig. 7G, H). CT angiography and MR venography of the brain were unremarkable. Transesophageal echocardiography did not show any high-risk cardiac source of embolic stroke.

Clinical Questions

- 1. What is the diagnosis in this case?
- 2. What is the underlying pathophysiology and etiology of this clinicoradiologic syndrome?
- 3. How should you treat a patient with this disorder?

Discussion

The differential diagnosis in this case includes ischemic stroke, cerebral venous thrombosis, hypertensive encephalopathy, PRES/reversible cerebral vasoconstrictive syndrome and eclampsia, and neoplasm. Taking into account the clinical presentation

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Fig. 7. (*A*, *B*) CT scan of the brain (axial sequence) shows multiple hypodensities involving the occipital lobes bilaterally. (*C–F*) MRIs of the brain (axial sequence) T2 (FLAIR) image show multiple hyperintensities involving the left cerebellar hemisphere (*C*), occipital lobes (*D–F*), and part of the frontoparietal lobes (*F*). (*G*, *H*) MRIs of the brain (axial sequence) DWI image (*G*) show an area of hyperintensity close to the parieto-occipital lobe on the right side and a corresponding area of low signal on apparent diffusion coefficient sequence (*H*). An arrow delineates the key neuroradiological feature referred to in the respective figure segment.

and imaging studies, the most likely diagnosis is PRES associated with eclampsia. The clinical features of PRES include headache, seizure, encephalopathy, and visual disturbance.

The neuroradiologic findings include focal reversible vasogenic edema that is best seen on brain MRI. However, PRES is a misnomer, as there can be involvement of the anterior circulation of the brain and gray matter. In addition, the neuroradiologic findings may be irreversible.

The precise underlying pathophysiology is not well defined. Rapidly developing hypertension leads to a breakdown in cerebral autoregulation. It is more pronounced in the posterior circulation of the brain, where there is a relative lack of a protective sympathetic innervation compared with the anterior brain circulation. Hyperperfusion leads to protein and fluid extravasation, which results in focal vasogenic edema. There is endothelial dysfunction in PRES patients with associated preeclampsia, eclampsia, and those who are treated with chemotherapeutic agents.^{21,25} PRES has been reported in patients with hypertensive encephalopathy, eclampsia, renal impairment, and toxicity related to various chemotherapeutic and immunosuppressive agents (eg, gemcitabine, carboplatin, ciclosporin), especially in patients with hematopoietic malignancies in whom high-dose chemotherapeutic agents are used.²⁵

Moreover, a strong association between eclampsia and the characteristic neuroradiologic findings of PRES have been reported. Furthermore, the symptoms of PRES in nonpregnant patients are similar to the prodromal symptoms of eclampsia. In one study, the incidence of PRES was more common among cases of postpartum eclampsias compared with antepartum eclampsias. Also, it was noted that PRES was more common in primigravidas and younger age group patients.²⁶ Furthermore, PRES and the seizures of eclampsia seem to be pathophysiologically related.²⁷

Early recognition of PRES is important for proper management and avoidance of complications.²⁸ In patients with PRES, immediate withdrawal of triggers, such as chemotherapeutic agents and rapid control of BP are recommended.²⁵ We are not aware of any large-scale clinical trials that address precise BP-lowering targets and time to reach the BP-lowering target in PRES. Based on one source, we believe that it is reasonable to decrease the mean arterial pressure by 20% to 25% within the first 2 hours and to bring the BP down to a target of 160/100 mm Hg within the first 6 hours. Furthermore, too rapid BP reduction is not recommended, as it might decrease the cerebral perfusion pressure resulting in cerebral hypoperfusion and ischemia.²⁷ In addition, a sudden decrease in BP may adversely affect uteroplacental blood flow; therefore, close maternal BP and fetal monitoring are recommended.^{29,30}

Prognosis is good in most of the cases of PRES; however, secondary complications, such as status epilepticus, intracranial hemorrhage, and large ischemic stroke can cause significant morbidity and mortality.^{28,29,31} Our patient did not have any long-term complications. She had complete recovery of focal deficits at the time of her 6-month follow-up visit.

SUMMARY

We provide a series of interesting neurologic cases from our practice and discuss diagnosis and treatment. As a number of the cases are uncommon causes of stroke, there is no high-level evidence base to guide the clinician in relation to diagnosis and treatment. Therefore, one must recognize the pattern of clinical features to arrive at the proper diagnostic workup and diagnosis and exercise good clinical judgment in relation to reasonable treatment. Careful neurologic history and examination remain paramount to determining the correct diagnosis and subsequent treatment.

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