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Case 34-2025: A 57-Year-Old Woman with Visual Disturbances and Right-Arm Shaking

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PRESENTATION OF CASE

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CME



Dr. Ashwin Amurthur (Neurology): A 57-year-old woman was admitted to this hospital because of persistent visual disturbances involving the right visual field, episodes of shaking of the right arm and leg, and hyperintensities on T1-weighted magnetic resonance imaging (MRI) of the head. ①

The patient had been in her usual state of health until 2 years before the current presentation, when frontotemporal and cervico-occipital headaches developed. The headaches occurred every 1 to 2 months, were associated with photophobia, and typically lasted for several hours to 1 day. ②

Nineteen months before the current presentation, the patient was evaluated by her primary care physician. The physical examination was normal. A diagnosis of migraines was considered. MRI of the head was performed, and T2-weighted fluid-attenuated inversion recovery (FLAIR) sequences revealed signal abnormalities in the subcortical and periventricular white matter, which were considered to be nonspecific. The patient was evaluated by a neurology consultant, who agreed with the diagnosis of migraines and recommended the use of intranasal zolmitriptan.

Six months before the current presentation, a severe headache developed suddenly while the patient was visiting a family member. She was evaluated at another hospital. Confusion was noted, along with generalized tonic-clonic seizure activity. ③

Dr. Shenghua Zhu: Computed tomography (CT) of the head revealed hyperdensity concentrated in the sulci of the left parietal lobe, a finding that was thought to be indicative of small-volume convexal subarachnoid hemorrhage (Fig. 1A). CT angiography and venography of the head and neck showed asymmetric prominence of the left distal branches of the anterior cerebral artery that extended to the left posterior parietal region, where the presumed subarachnoid hemorrhage was located, as well as mild prominence of the cortical venous structures in that region — findings consistent with a pial arteriovenous fistula (Fig. 1B, 1C, and 1D). MRI of the head was performed 10 hours after the initial presentation. T1-weighted sequences showed intrinsic hyperintensities that were most prominent along the left parietal lobe, ④ ⑤

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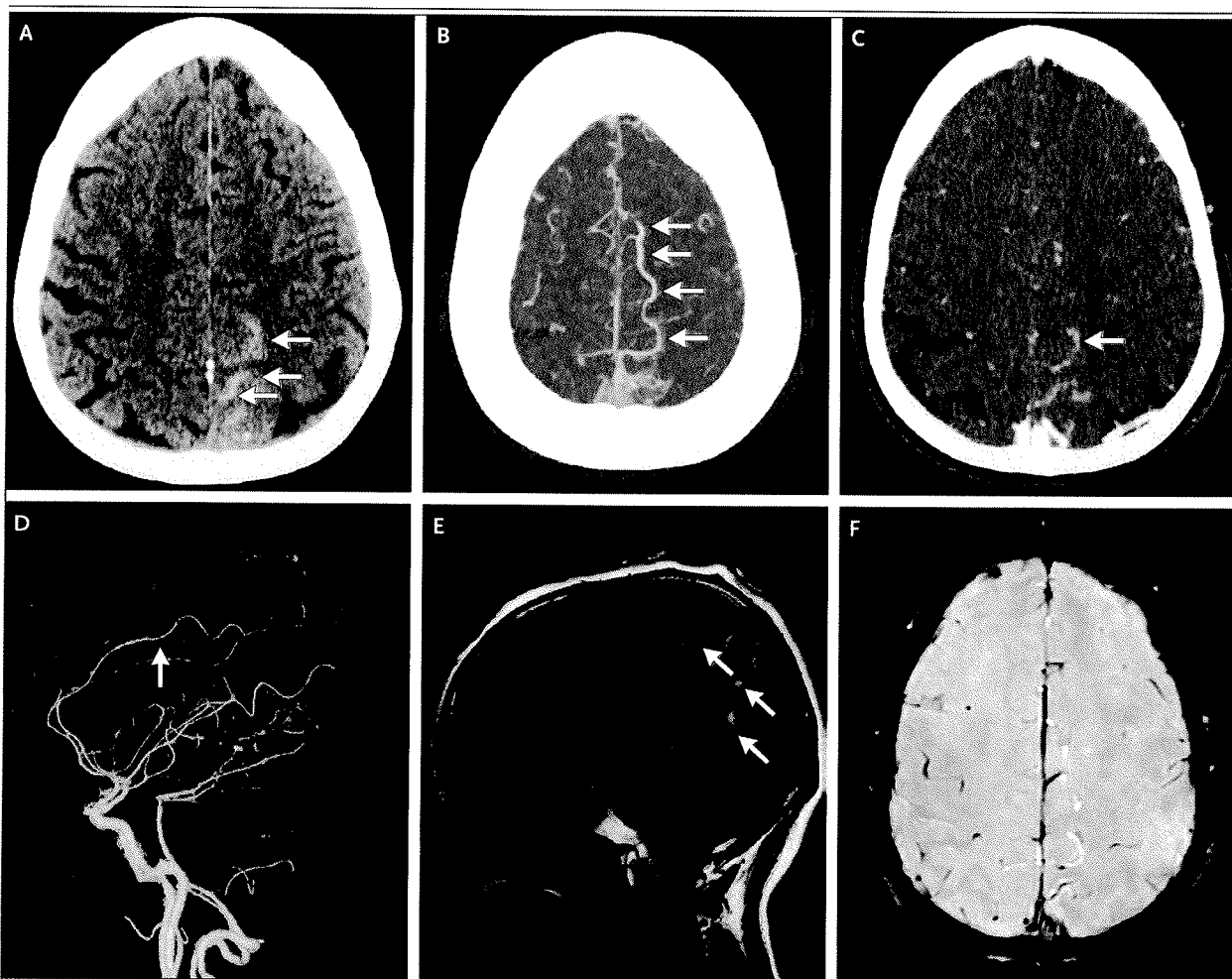


Figure 1. Imaging Studies of the Head, 6 Months before the Current Presentation.

An axial CT image (Panel A) shows hyperdensity concentrated in the sulci of the left parietal lobe (arrows). Axial images from CT angiography and venography (Panels B and C, respectively) show asymmetric prominence of the left distal branches of the anterior cerebral artery (Panel B, arrows) and mild prominence of the cortical veins (Panel C, arrow) in the paracentral parietal lobe. A three-dimensional reformat image from CT angiography (Panel D) also shows asymmetric prominence of the left anterior cerebral artery (arrow). A sagittal T1-weighted MRI (Panel E) shows intrinsic hyperintensities in the sulci of the left parietal lobe (arrows). An axial susceptibility-weighted angiographic MRI (Panel F) shows no corresponding susceptibility signal in this region.

a finding consistent with the presence of blood products related to subarachnoid hemorrhage (Fig. 1E and 1F).]

⑥ Dr. Amurthur: Cerebral angiography reportedly revealed a fistula in the left middle meningeal artery that was connected to the superior sagittal sinus. An embolic agent was injected into the fistula, and follow-up angiography showed successful complete occlusion of the fistula. After the procedure, the patient continued to have headaches, and visual disturbances developed, including visual hallucinations and double vision that predominantly occurred on the right side, as well as visual

auras. Treatment with lacosamide was started for possible seizures, and visual symptoms abated.]

After discharge from the other hospital, the patient returned home and attended a follow-up visit in the neurology clinic affiliated with this hospital. Treatment with lacosamide was continued, and in consultation with the neurosurgery team, plans were made for repeat cerebral angiography to be performed in 6 months.]

One month later, and 5 months before the current presentation, generalized tonic-clonic seizure activity recurred. The patient was admitted to this hospital.

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Dr. Zhu: CT of the head showed hyperattenuating embolic material with a linear branching pattern along the falx cerebri and cortical vessels surrounding the posterior superior sagittal sinus in the left cerebral hemisphere, findings suggestive of previous embolization of a vascular malformation (Fig. 2A). The area where the presumed subarachnoid hemorrhage had been noted previously was stable on both CT and MRI (Fig. 2B and 2C).

10

Dr. Amurthur: Cerebral angiography identified evidence of previous embolization but showed no evidence of a persistent shunt or venous thrombosis. The patient reported persistent visual disturbances, which did not have a correlate on

electroencephalography (EEG) and were suspected to be due to the presence of blood in the visual cortex.

11

Dr. Zhu: MRI of the head was also performed and revealed several foci of diffusion restriction in the right parietal lobe on diffusion-weighted sequences that corresponded to hyperintensity on FLAIR sequences — findings compatible with acute or early subacute ischemic infarcts, which were thought to be embolic (Fig. 2D, 2E, and 2F). Additional, nonspecific hyperintensities in the periventricular white matter were unchanged from the findings seen on MRI 18 months earlier.

Dr. Amurthur: The dose of lacosamide was in-

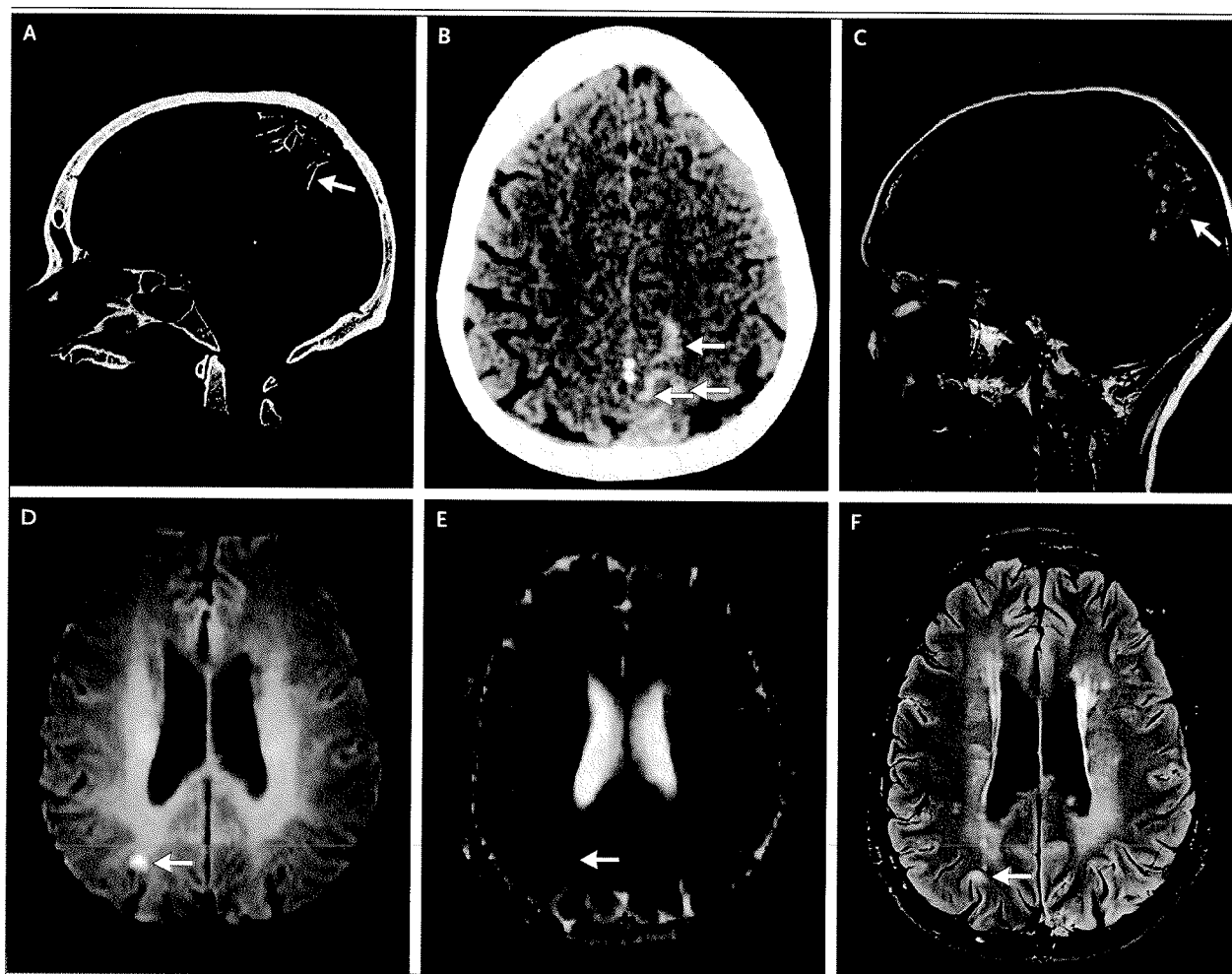


Figure 2. Imaging Studies of the Head, 5 Months before the Current Presentation.

A sagittal CT image (Panel A) shows hyperattenuating embolic material with a linear branching pattern along the falx cerebri (arrow). An axial CT image (Panel B) and a sagittal T1-weighted MRI (Panel C) show stability of the presumed subarachnoid hemorrhage (arrows). A punctate focus of hyperintensity in the right parietal lobe on an axial diffusion-weighted MRI (Panel D, arrow) corresponds to hypointensity on an axial apparent-diffusion-coefficient MRI (Panel E, arrow) and to hyperintensity on an axial fluid-attenuated inversion recovery MRI (Panel F, arrow).

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creased, and the patient was discharged from the hospital.

One month later, and 4 months before the current presentation, the patient sustained a fall during an episode of dizziness and lightheadedness, and she was again admitted to this hospital. She had persistent visual disturbances involving the right visual field that she described as "flashing lights."

Dr. Zhu: CT of the head again showed hyperdensity in the sulci of the posterior paracentral lobule in the left parietal lobe and, to a lesser degree, along the precentral sulcus of the right frontal lobe. No radiologically significant change from the previous study was noted.

Dr. Amurthur: Long-term EEG monitoring showed no evidence of seizure activity or an EEG correlate that would explain the visual disturbances. Examination by an ophthalmology consultant was normal. The patient's presenting symptoms were thought to be due to presyncope, and no changes were made to the antiepileptic regimen.

One month later, and 3 months before the current presentation, blurry vision in both eyes and hoarseness developed. The patient had another generalized tonic-clonic seizure, and she was again admitted to this hospital.

Dr. Zhu: CT of the head showed a stable appearance of the hyperattenuation involving the sulci of the left parietal lobe, as well as a few scattered foci along the sulci of the right frontal lobe. These findings were thought to be consistent with previous embolization of the fistula. MRI of the head was also performed, and T1-weighted sequences showed nodular intrinsic hyperintensities in the sulci and leptomeninges of the left parietal and occipital lobes, with associated susceptibility signals that were suggestive of mineralization. These findings were similar to the MRI findings seen 2 months earlier. New nodular intrinsic hyperintensities were noted along the dorsal midbrain and in the posterior right sylvian fissure, the operculum in the right frontal lobe, the convexity of the right anterior temporal lobe, and the convexity of the left and right medial frontal lobes.

Dr. Amurthur: Lumbar puncture was performed, and cerebrospinal fluid (CSF) analysis showed 1 red cell per microliter, 1 nucleated cell per microliter, and normal levels of glucose and protein. Positron-emission tomography of the head showed low fluorodeoxyglucose uptake in the region where the left parieto-occipital sulcal abnormalities had

been seen on recent MRI, which indicated hypometabolism in that area. Treatment with levetiracetam was added, and the patient was discharged from the hospital. Cytologic evaluation of the CSF showed no malignant cells, and flow cytometry of the CSF showed no evidence of a monoclonal B-cell population.

One month later, and 2 months before the current presentation, the patient was evaluated in the otolaryngology clinic affiliated with this hospital because of hoarseness. Flexible laryngoscopy revealed immobility of the right vocal cord. The patient was also evaluated in the neurology clinic affiliated with this hospital because of increased sleepiness and episodes of tremulousness in the right arm and leg. Given the possibility of medication side effects, the course of levetiracetam was tapered off, and treatment with clobazam was started. The episodes of tremulousness became less frequent but did not abate, and the visual disturbances involving the right visual field persisted.

On the day of the current presentation, the episodes of tremulousness in the right arm and leg increased in frequency and length, lasting up to 10 minutes at a time. No loss of consciousness occurred. The patient presented to the emergency department of this hospital for evaluation.

The patient reported increasingly frequent visual disturbances involving the right visual field, which she described as "seeing rainbows, circles, and flashes." Additional medical history included allergic rhinitis, vitamin D deficiency, and migraines. Medications included clobazam and lacosamide. She had no known allergies to medications. Before this illness, the patient had worked in real estate. She lived with her husband and two children in Massachusetts. She was a lifelong non-smoker, consumed alcohol only occasionally, and did not use illicit drugs. Her family history included migraines in her father and brother, stroke in her mother, and melanoma in her father.

On examination, the temporal temperature was 36.5°C, the blood pressure 104/62 mm Hg, the heart rate 57 beats per minute, the respiratory rate 18 breaths per minute, and the oxygen saturation 96% while the patient was breathing ambient air. The body-mass index (the weight in kilograms divided by the square of the height in meters) was 18.5. The patient was alert and oriented. Language was fluent, and comprehension, repetition, and naming were intact. On examination of the cranial