Burnt-out arteriovenous malformation mimicking unilateral Fahr's disease: a diagnostic pitfall in adult-onset seizure: a case report

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European Journal of Medical Case Reports
Volume XX(XX):01–07
DOI: XXXX

ABSTRACT

Background: Intracranial calcifications are a common radiological finding, but their significance varies widely depending on the pattern and distribution. While Fahr's disease is often considered in such cases, it typically presents with bilateral and symmetrical involvement. Unilateral, extensive calcifications, especially when associated with cerebral atrophy, should prompt a broader differential.

Case Presentation: We describe a 43-year-old man with no significant medical history who presented with his first-ever seizure. He was found unresponsive by his wife, with tongue biting and post-ictal confusion. By the time he arrived in the emergency department, he had returned to baseline, and his neurological examination was completely normal. Initial computed tomography (CT) imaging revealed extensive right-sided subcortical calcifications with associated cerebral atrophy, raising the possibility of Fahr's syndrome. However, further imaging revealed a different etiology. Magnetic resonance imaging indicated dilated convoluted signal-void formations with post-contrast enhancement, and Susceptibility-weighted imaging confirmed widespread blooming matching the calcifications. At the right Middle Cerebral Artery (MCA) bifurcation, CT angiography revealed a 4.1 mm focal dilatation with surrounding tortuous arteries. Ultimately, digital subtraction angiography did not demonstrate an active arteriovenous malformation (AVM) but revealed an ectatic MCA bifurcation with a small aneurysmal bulge. Taken together, the findings were most consistent with chronic changes from a previously existing, now-regressed vascular malformation.

Conclusion: This case highlights how unilateral calcifications can be misleading and emphasizes the importance of correlating imaging patterns with clinical context. A vascular etiology, including a probable "burnt-out" AVM, should be considered in similar presentations to avoid misdiagnosis.

Keywords: Arteriovenous Malformation, Burnt-out AVM, Fahr's Disease, Intracranial calcification, Unilateral cerebral calcification.

Type of Article: Case Report **Specialty:** Neurology

Received: 15 June 2026

Revised (1): 30 August 2026

Accepted: 31 August 2026

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Background

Intracranial calcifications are calcium deposits within the brain parenchyma or vasculature. They can be divided into several major categories: physiologic, dystrophic, congenital disorders, infectious, vascular, neoplastic, metabolic, inflammatory, and toxic diseases [1]. Their prevalence varies from 1% in young people to 38% in the elderly. Intracranial calcification can be primary, as observed in several early- and late-onset genetic syndromes, or secondary to systemic alterations of phosphate-calcium metabolism, intrauterine and postnatal infections, hypoxic-ischemic injuries, toxic exposures, brain tumors, and autoimmune disorders. Among these diseases, Fahr's disease is the most common neurological disorder associated with late-onset brain calcification [2].

Fahr's disease is a rare neurological illness characterized by aberrant idiopathic calcification of the basal ganglia, which is typically inherited autosomally [3]. Notably, although Fahr's disease and Fahr's syndrome are still commonly used in clinical settings, the term primary familial brain calcification (PFBC) is preferred in current nomenclature. It is clinically defined by the varying occurrence of movement disorders, recurrent headaches, and psychiatric indications in combination with bilateral presence of intracranial calcifications, usually in the basal ganglia, but can also involve other locations in the brain [2,4]. The anatomical position, distribution, size, and shape are significant findings that, when combined with the clinical history and age group, can aid in the differential diagnosis. Unilateral intracranial calcifications are infrequent and

typically imply a pathogenic process rather than a physiological or systemic condition. In contrast to the more common bilateral patterns seen in certain metabolic or genetic disorders, asymmetric or focal calcifications warrant a more comprehensive differential diagnosis. Recognizing this distinction is vital since the distribution and pattern of calcification guide further diagnostic examination and help narrow down the underlying cause [5].

Brain arteriovenous malformations (AVMs) are aberrant connections between arteries and veins that cause arteriovenous shunting through an intervening network of vessels known as the nidus. AVMs are the second most frequent vascular malformation in the brain, as well as the second most common calcified brain lesion. Calcification in arteriovenous malformations is uncommon, but it can occur in chronic lesions, most likely due to long-term venous hypertension, repetitive microhemorrhage, or dystrophic alterations in ischemic parenchyma [6].

In rare cases, cerebral arteriovenous malformations can spontaneously thrombose and regress, resulting in loss of angiographic shunting and the presence of residual alterations such as calcification, gliosis, or localized atrophy. This condition, known as a "burnt-out" AVM, can complicate radiological interpretation, especially when active vascular components are no longer visible [7]. Computed tomography (CT) or magnetic resonance imaging (MRI) is used to detect brain AVMs. A combination of MRI and angiography is used to plan therapy and predict the prognosis and related hazards of surgical, endovascular, or radiological therapy. Management is determined by patient age and risk profile; invasive methods are examined for younger high-risk individuals, whereas conservative medical management is chosen in older patients lacking high-risk traits [8].

This case illustrates a diagnostic challenge in which unilateral calcifications first suggested a primary neurodegenerative condition, such as Fahr's disease, but additional investigation found an underlying regressed vascular malformation.

Case Presentation

A 43-year-old Indian male was brought to the emergency department after an episode of sudden loss of consciousness at home. According to his wife, he had just woken up and was attempting to get out of bed when he suddenly became unresponsive. The episode lasted a few minutes and was associated with tongue biting, snoring-type breathing, and confusion afterward. There was no reported incontinence. By the time emergency medical services arrived, he had begun to regain consciousness but had no recollection of the event. Routine laboratory investigations were within normal limits, with no evidence of a metabolic cause for intracranial calcification (Tables 1 and 2).

A non-contrast CT scan of the brain revealed extensive calcifications involving the right frontal and parietal subcortical regions, along with noticeable atrophy of the right cerebral hemisphere (Figures 1,2 and 3). The asymmetry was striking and immediately raised questions about the underlying cause.

Given these findings, an MRI was performed for further characterization (Figures 4 and 5). Susceptibility-weighted

Table 1. Blood chemistry.

PARAMETER	DAY 2	DAY 1	REFERENCE RANGE
Urea (mmol/l)	3.8	3.5	2.5-7.1
Creatinine (µmol/l)	92	97	60-110
Sodium (mmol/l)	144	138	135-145
Potassium (mmol/l)	3.9	4.0	3.5-5.0
Chloride (mmol/l)	105	100	98-107
Bicarbonate (mmol/l)	26	24	22-28
Phosphorus (mmol/l)	1.24	_	0.8-1.5
Calcium (mmol/l)	_	2.21	2.15–2.55
Adjusted Calcium (mmol/l)	_	2.27	2.15–2.55
Magnesium (mmol/l)	0.86	_	0.7-1.0
Total Bilirubin (µmol/l)	_	12	5-21
Total Protein (g/l)	_	69	60-80
Albumin (g/l)	_	41	35-50
ALP (U/l)	_	58	44-147
ALT (U/l)	_	20	7-56
AST (U/l)	_	24	10-40
Random Glucose (mmol/l)	_	5.5	4.0-7.8
CRP (mg/l)	_	0.9	<5
Lactate (mmol/l)	0.9	_	0.5-2.2

Table 2. Endocrinology.

PARAMETER	VALUE	REFERENCE RANGE
Vitamin D (ng/ml)	9	20-50
Parathyroid Hormone (pg/ml)	54	15-65

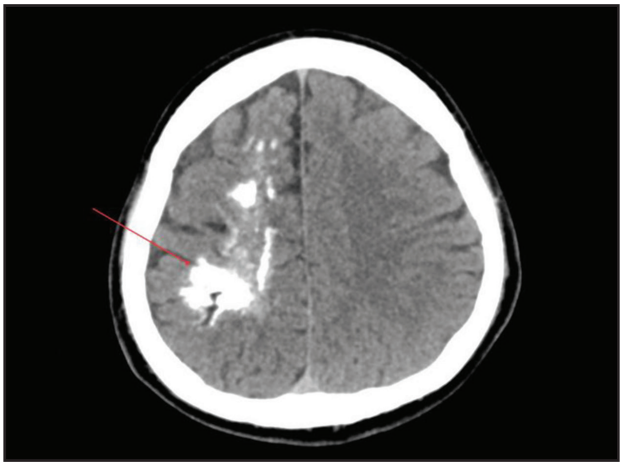

Figure 1. *Non-contrast axial CT brain demonstrating extensive unilateral gyriform and nodular cortical-subcortical calcifications involving the right frontal and parietal lobes (red arrows), with a linear calcified component extending along the periventricular region.*

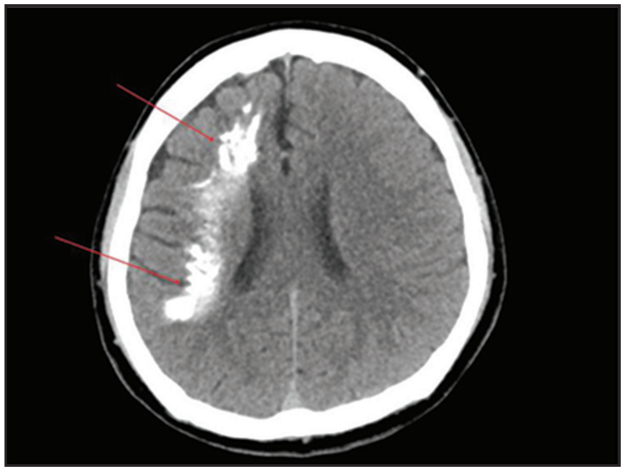

Figure 2. *Axial non-contrast CT brain showing dense right parietal cortical-subcortical calcified lesions with surrounding chronic parenchymal changes and adjacent linear calcification extending superiorly (red arrow).*

imaging (SWI) revealed widespread bizarre-shaped blooming in the right frontal white matter, similar to the CT calcifications, whereas the basal ganglia were noticeably spared. A cluster of dilated tortuous signal-void structures was found in the right frontal subcortical area at the level of the frontal horn on T1 post-contrast sequences, indicating patchy post-contrast enhancement. With a pronounced right lateral ventricle, cortical sulci, Sylvian fissure, and CSF gaps across the frontal convexity, the right cerebral hemisphere appeared smaller than the left, indicating right cerebral atrophy. Strong suspicion of an underlying vascular abnormality was raised by MR

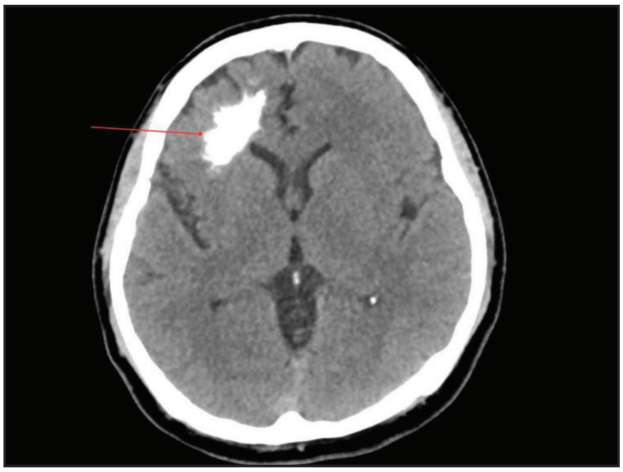

Figure 3. *Axial CT brain image demonstrating a large right frontal parasagittal calcified lesion without significant surrounding edema or midline shift (red arrow).*

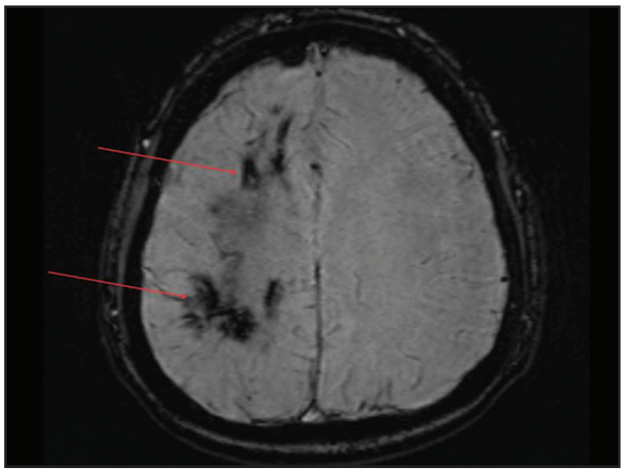

Figure 4. *Susceptibility-weighted MRI (SWI) showing marked blooming artifacts corresponding to the heavily calcified lesions in the right frontal and parietal regions (red arrows).*

angiography, which showed asymmetric dilatation of the right Middle Cerebral Artery (MCA) Sylvian branches relative to the left (Figure 6). Digital subtraction angiography (DSA) was necessary because CT angiography showed a 4.1-mm focal dilatation at the right distal M1 MCA bifurcation with surrounding tortuous arteries; an active AVM could not be completely ruled out.

Digital subtraction angiography was carried out using a right femoral puncture, selective catheterization of the left vertebral artery, internal and external carotid arteries, and standard Anteroposterior (AP), lateral, and oblique projections with rotational angiography and 3D reconstruction of the right Internal Carotid Artery (ICA) to clarify the vascular anatomy. There were no aberrant feeding arteries, early draining veins, residual nidus, or active arteriovenous malformations. An ectatic right MCA bifurcation with a 3-mm aneurysmal bulge was shown in the investigation. The route and caliber of all other cerebral arteries, including the left middle cerebral artery, anterior cerebral artery, and bilateral common carotid arteries,

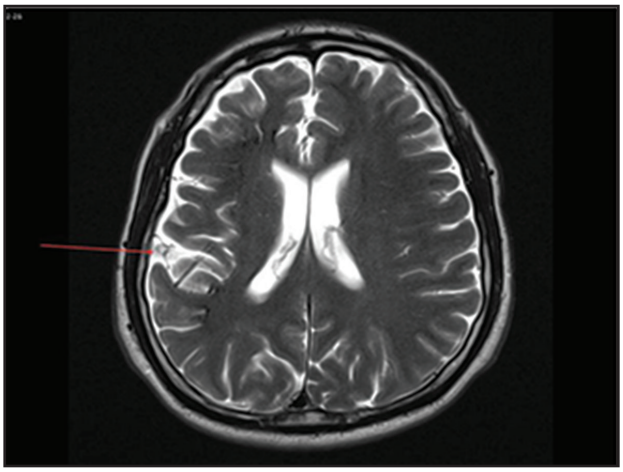

Figure 5. *Axial T2-weighted MRI demonstrating focal encephalomalacic and gliotic changes adjacent to the right parietal calcified lesion with associated cortical volume loss (red arrow).*

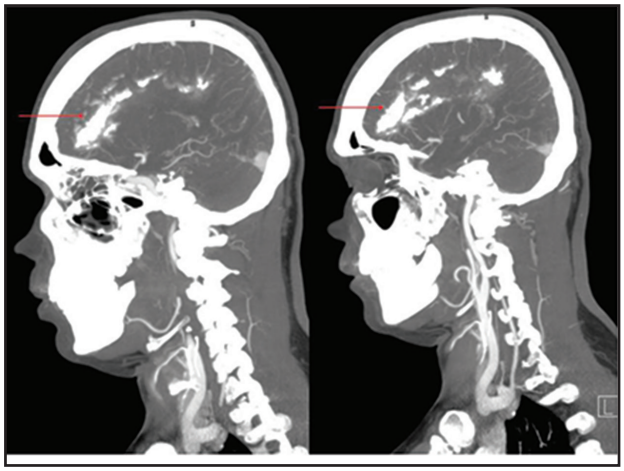

Figure 6. *Sagittal CT angiographic reconstructions demonstrating elongated dense calcified lesions along the right frontal-parietal convexity without evidence of a definite enhancing vascular nidus or arteriovenous shunting (red arrows).*

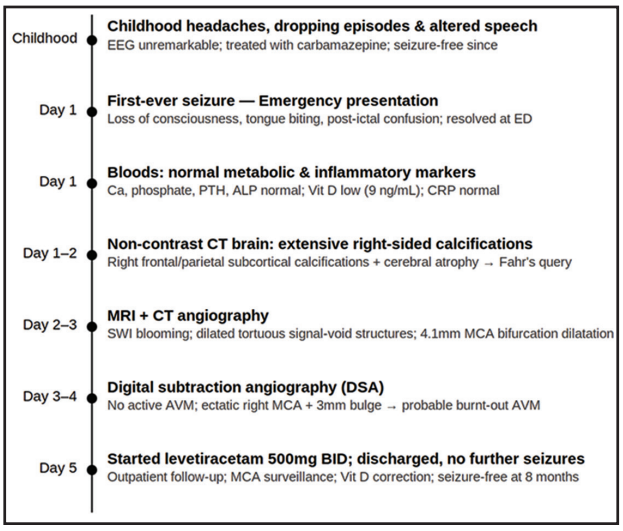

Figure 7. *Clinical timeline illustrating the diagnostic workup of a 43-year-old man presenting with first-ever seizure, ultimately found to have findings consistent with a probable burnt-out arteriovenous malformation.*

were normal. Considering these findings, the overall picture was felt to represent chronic sequelae of a previously existing vascular lesion, most likely a regressed or thrombosed AVM. The patient was started on levetiracetam and remained clinically stable, with no further seizures during his hospital stay. He was discharged on levetiracetam 500 mg twice daily. Given the underlying structural etiology, long-term antiseizure medication was maintained at the outpatient neurology follow-up. The patient was taking 500 mg of levetiracetam twice a day and reported no further seizures at follow-up outpatient neurology visits 2 and 6 months after presentation. He was still seizure-free at the time of final contact, which was about 8 months after the index episode. The chronological progression of investigations is summarized in Figure 7.

Discussion

Extensive unilateral cerebral calcification with a new-onset seizure in a middle-aged man is an unusual combination, and the initial CT report raising Fahr-type calcification syndrome as the leading possibility was not unreasonable. Fahr's disease, now termed PFBC, is idiopathic/genetic, while Fahr's syndrome denotes an identical pattern from a recognized metabolic, endocrine, infectious, or toxic cause [3,4]. PFBC commonly presents in the fourth or fifth decade with neurological symptoms, and brain CT is the primary modality for its diagnosis [9]. Two features set this case apart from either. The calcification was strictly unilateral; both PFBC and Fahr's syndrome require bilateral and symmetric basal ganglia involvement as a diagnostic prerequisite [9,10]. Additionally, the deposits were confined to the right frontal and parietal subcortical white matter, entirely distinct from the basal ganglia, thalamus, and dentate nucleus that PFBC and Fahr's syndrome characteristically involve [11]. The full metabolic workup was normal: calcium, phosphorus, PTH, magnesium, and alkaline phosphatase were within reference ranges throughout, effectively ruling out hypoparathyroidism and other biochemical derangements that drive Fahr's syndrome and other forms of secondary brain calcification [9,10]. Vitamin D was deficient (9 ng/ml), but without any accompanying calcium-phosphate disturbance, it cannot account for extensive unilateral subcortical calcium deposition. Cavernous malformation, calcified tumor, post-infectious alterations, and developmental venous abnormality were among the other reasons for unilateral calcification that were taken into consideration but ruled out due to negative angiographic findings, lack of mass effect, and imaging morphology [1,5]. In the setting of unilateral calcification with cerebral atrophy, the combination of dilated tortuous vascular structures on MRI, asymmetric MCA dilatation on MRA, and ectatic MCA bifurcation on DSA supported the most logical diagnosis of a likely spontaneously regressed vascular abnormality.

Calcification in AVMs arises through chronic venous ischemia, repeated microhemorrhage, intraluminal thrombosis, and dystrophic deposition in ischemic parenchyma caused by vascular steal [5]. Seizures occur in 20%-45% of patients with cerebral AVMs, and cortical or subcortical locations are particularly epileptogenic [12]. The patient's early neurological symptoms, which included headaches, occurrences of dropping objects, and sporadic altered speech, were investigated with Electroencephalogram (EEG), which was unremarkable, and managed with carbamazepine, discontinued at age seven. In rare cases, cerebral AVMs undergo spontaneous thrombosis and angiographic obliteration, typically around 26 months after a precipitating event such as hemorrhage [13]. As the AVM involutes and regresses, it becomes sclerotic, calcified, and surrounded by hemosiderin-stained and gliotic parenchyma, leaving inert residue with no active shunting [5]. This end state, commonly called a "burnt-out" AVM, carries genuine diagnostic risk; on CT alone, it is indistinguishable from PFBC or Fahr's syndrome, as this case clearly demonstrates.

The sequential imaging in this case illustrates why CT alone is inadequate when unilateral calcification is encountered. MRI showed dilated convoluted signal-void structures with post-contrast enhancement, and SWI verified widespread blooming associated with the calcifications. At the right MCA bifurcation, CT angiography revealed a 4.1-mm focal dilatation with surrounding tortuous arteries. Digital subtraction angiography then provided definitive clarification. Selective catheterization with rotational 3D reconstruction of the right ICA confirmed the absence of active arteriovenous shunting, findings consistent with spontaneous obliteration of a previously existing vascular malformation. DSA remains the gold standard for this determination: it offers spatial and temporal resolution that non-invasive techniques cannot match, and it is the only modality currently capable of definitively confirming the absence of residual arteriovenous shunting [14].

With an active AVM excluded, management was directed at seizure control. Levetiracetam was initiated, a choice consistent with current evidence supporting antiseizure drug therapy as the primary approach to AVM-related epilepsy [12]. Because there was a structural epileptogenic lesion in the right frontal region, long-term antiseizure therapy was considered acceptable. Two EEGs were conducted in the outpatient neurology clinic after discharge, and both showed typical awake findings with no recorded epileptiform activity. Vascular intervention was not indicated, given the negative DSA. A 4.1-mm focal dilatation at the right distal M1 segment of the MCA at the bifurcation was seen on CT angiography. This finding suggested a tiny aneurysm rather than focal vascular ectasia, given the mild irregularity and the surrounding tortuous arteries in the Sylvian fissure. A 3-mm aneurysmal bulge at the ectatic right MCA bifurcation was verified by a subsequent DSA. This discovery is more consistent with focal vascular ectasia than a true saccular aneurysm, given the imaging features and the probable hemodynamic alterations linked to a prior vascular abnormality. The treating neurovascular team has advised yearly periodic neurovascular imaging surveillance. Vitamin D deficiency should be corrected; while it did not cause the calcification here, sustained hypovitaminosis D has independent neurological implications. Spontaneous AVM regression is not always permanent: recanalization has been documented years after confirmed angiographic obliteration and can culminate in rupture, making continued imaging follow-up necessary [15].

Conclusion

This case report demonstrates how readily unilateral intracranial calcifications can be misconstrued, especially when a seizure is occurring for the first time. Fahr's disease may be considered at first, but its diagnosis depends on symmetrical and bilateral damage, which was absent in this case. Based on available literature, even in the absence of active angiographic shunting, a previously existent but now regressed (or "burnt-out") arteriovenous malformation may manifest in this manner. This case underscores how crucial it is to match imaging patterns to the clinical context and, when the presentation is unusual, to pursue further evaluation with MRI and digital subtraction angiography.

What is New?

Burnt-out AVMs can closely mimic Fahr's disease on CT imaging, particularly when calcifications are unilateral and extensive. A normal calcium-phosphate-PTH axis should redirect the clinician toward a vascular etiology. Multimodal imaging including MRI with SWI and DSA is essential to confirm spontaneous AVM regression and exclude active vascular disease.

List of Abbreviations

AVM	Arteriovenous Malformation
CRP	C-Reactive Protein
CT	Computed Tomography
CTA	Computed Tomography Angiography
DSA	Digital Subtraction Angiography
ICA	Internal Carotid Artery
MCA	Middle Cerebral Artery
MRI	Magnetic Resonance Imaging
PFBC	Primary familial brain calcification
PTH	Parathyroid Hormone
SWI	Susceptibility-Weighted Imaging

Conflicts of interest

The authors declare that they have no conflicts of interest regarding the publication of this case report.

Patient public involvement

Patient and Public Involvement (PPI): No patients or members of the public were involved in the design, conduct, reporting, or dissemination plans of this research.

Consent to publication

Written informed consent was obtained from the patient for publication of this case report and all accompanying clinical images, including non-contrast CT brain, MRI with contrast, CT angiography, and digital subtraction angiography.

Availability of data and materials

All data are included in the manuscript. For further discussion regarding this report, please contact the corresponding author.

Funding

Not applicable.

Ethical approval

Ethical approval was granted by the Medical Research Center, Hamad Medical Corporation, Doha, Qatar (Reference No. MRC-04-26-150).

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Summary of the case

1	Patient (gender, age)	43 years, Male
2	Final diagnosis	Probable Burnt-out arteriovenous malformation
3	Symptoms	First-ever seizure with loss of consciousness, tongue biting, post-ictal confusion
4	Medications	Levetiracetam
5	Clinical procedure	CT brain, MRI with SWI, CT angiography, Digital Subtraction Angiography
6	Specialty	Neurology