Idiopathic De Novo Arteriovenous Malformation: A Rare Acquired Intracranial Lesion

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Abstract

The de novo development of cerebral arteriovenous malformations (AVMs) in adults is an exceedingly rare event that has prompted the theory that a “second hit” is required to induce AVM formation. The authors document development of an occipital AVM in an adult a decade and a half after a brain magnetic resonance imaging (MRI) disclosed no abnormality. A 31-year-old male with a family history of AVMs and a 14-year history of migraines with visual auras and seizures presented to our service. Because of the onset of a first seizure and migraine headaches at age 17, the patient underwent high-resolution MRI that showed no intracranial lesion. After 14 years of progressively worsening symptoms, he underwent a repeat MRI that demonstrated a new de novo Spetzler-Martin grade 3 left occipital AVM. The patient received anticonvulsants and underwent Gamma Knife radiosurgery for his AVM. This case suggests that patients with seizures or persistent migraine headaches should have periodic repeat neuroimaging to exclude the development of a vascular cause despite an initial negative MRI.

Keywords
► arteriovenous malformations
► Gamma Knife radiosurgery
► de novo
► genetic
► seizures

Introduction

Cerebral arteriovenous malformations (AVMs) are rare, complex vascular lesions of uncertain pathogenesis involving dysplastic vessel formation.1 Originally thought to be exclusively congenital,2 intracerebral AVMs are now known to also arise de novo3–5 in those with genetically predisposed disorders (e.g., ataxia telangiectasia).6 Some authors suggest that AVMs arise when individuals with genetic predispositions experience one or more “second hits.”7 Such “second hits” include events that promote dysplastic angiogenesis such as brain hemorrhage, trauma, or venous sinus thrombosis.5 To the best of our knowledge, distinct “second hits” preceded all reported intracranial AVMs but one.5,8 Here, the authors present a rare case of a patient with a family history of AVMs presenting with an idiopathic de novo occipital AVM with no identifiable antecedent “second hit” and absence of any intracranial lesion on adolescence high-resolution imaging.
Case History

A 17-year-old male with a family history of AVM (including a brother with an AVM of the corpus callosum) presented to the emergency room due to migraines accompanied by visual auras and generalized seizure. The patient underwent high-resolution magnetic resonance imaging (MRI), which failed to demonstrate any intracranial pathology (Fig. 1A–C). The patient continued to have rare repeat seizures, but antiepileptic drugs were not initiated at any point due to the patient’s and parents’ preference. The patient had a history of prior intravenous drug use, hepatitis C infection, and pre-diabetes.

Fourteen years after his initial presentation, the 31-year-old patient reported to the emergency department for a head injury sustained during a seizure. Computed tomography angiography of the head revealed a 1.9 x 1.4 cm hyperattenuating vascular mass within the left occipital lobe (Fig. 2) with no evidence of hemorrhage. Cerebral angiography confirmed the presence of an unruptured 2.9 cm Spetzler-Martin grade 3 left occipital AVM (Fig. 3). The AVM was supplied by the calcarine and posterior temporal branches of the posterior cerebral artery and had both superficial and deep venous drainage. This occipital AVM was also demonstrated on MR imaging performed at that time (Fig. 1D–F).

Following diagnosis, the patient was started on levetiracetam and referred to our service for further evaluation. The patient subsequently underwent Gamma Knife radiosurgery, during which the margin dose of the AVM was 20 Gy, with 85% of the AVM receiving over 22 Gy, and the 12 Gy volume...
Documented de novo intracranial AVM formation is an exceedingly rare entity. We suggest that additional long-standing chronic conditions could act as comorbidity facilitators of AVM formation. In the context of persistent clinical symptoms such as migraine and seizures, repeat brain imaging is important to exclude delayed development of a treatable and high-risk AVM.

Authors’ Contributions
Tritan Plute was responsible for data collection, writing and primary generation of the manuscript. Prateek...
Agarwal was the primary resident involved with the case and provided insight into the case and assisted with writing. Aneek Patel was involved with the case write-up and provided neurosurgical insight. Arka Mallela provided key insight into the research process and assisted with manuscript writing. Lunsford was responsible for identifying the case as novel, obtaining patient consent, writing, and providing expert opinion regarding the case. Abou-Al-Shaar was responsible for the design of the report and advising Plute during the research process; he was also responsible for writing manuscript generation.

Ethical Approval
All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee of University of Pittsburgh Medical Center and with the 1964 Helsinki declaration and its later amendments. Informed consent was obtained from the patient included in the study.

Funding
None.

Conflict of Interest
None declared.

References
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