Anterior Cranial Fossa Dural Arteriovenous Fistula Presenting as Recurrent Multifocal Lobar Intracerebral Hemorrhages: A Case Report

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Abstract

We report a unique case and presentation of a 77-year-old man with atrial fibrillation who presented with complex imaging features, including bifrontal cerebral hematomas that progressed to multifocal macro- and microhemorrhages, vasogenic edema, and cortical/meningeal enhancement. Cerebrospinal fluid analysis was only remarkable for a protein elevation of 65 mg/dL. Leptomeningeal vessel biopsy failed to demonstrate evidence of amyloid angiopathy but did show concentric obstructive intimal fibroplasia, suggestive of chronic venous congestion. Conventional cerebral angiography showed a Cognard type IV dural arteriovenous fistulae fed by bilateral ethmoidal arteries draining directly into ectatic cortical veins. A chronic occlusion of the anterior third of the superior sagittal sinus was also noted. The fistula was successfully surgically obliterated via an uncomplicated left frontal craniotomy approach. After rehabilitation therapy, he was discharged home with a modified Rankin Scale score of 3. Because effective treatment exists, this case underscores the importance of identifying this rare etiology of intracerebral hemorrhage and pursuing adequate workup.

Keywords

dural arteriovenous fistula, anterior cranial fossa, arteriovenous malformation, vascular malformation, intracerebral hemorrhage

Introduction

Dural arteriovenous fistulae (dAVF) are pathologic shunts that directly connect arterial and venous circulations and account for 14% of cerebral arteriovenous malformations.¹ They most commonly involve the cavernous, sigmoid, and transverse sinuses,^{2,3} but 4% of dAVF are located in the anterior cranial fossa (ACF).¹ Features such as cortical venous drainage, common to ACF-dAVF, portend increased morbidity and mortality.⁴ Despite their high rate of hemorrhage (ranging 62%-91%),^{4,5} ACF-dAVF remain a treatable subgroup of dAVF. Treatment approaches include open surgery, endovascular obliteration, stereotactic radiosurgery, or a combination of these.⁶ The morbidity of dAVF hemorrhage can be substantial, and many patients do not survive or recover to a condition suitable for intervention.²

Case Description

A 77-year-old man with a history of hypertension, parkinsonism, and atrial fibrillation on aspirin and apixaban presented with headache, confusion, and cognitive changes. The patient

and wife denied any trauma; however, the patient had a small right occipital scalp hematoma and National Institutes of Health Stroke Scale (NIHSS) score was 0. Noncontrast head computed tomography (CT) revealed multifocal bifrontal intracerebral hemorrhages (2.7, 2.0, and 0.2 mL; intracerebral hemorrhage (ICH) score: 0; secondary ICH (sICH) score: 1; Figure 1). After reversal of anticoagulation with prothrombin complex concentrate, he was admitted to the neurosciences intensive care unit. Twelve hours after admission, his clinical course was complicated by right frontal hematoma expansion from 0.2 to 10.7 mL, (Figure 1) presenting as a generalized tonic-clonic seizure. Initial brain magnetic resonance imaging

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Figure 1. Computed tomography of bifrontal intracerebral hematomas. Noncontrast computed tomography of the brain demonstrates multifocal cerebral hematomas in the right medial orbital gyrus (A, arrow) and throughout left frontal lobe (B, arrows). Interval expansion of the right frontal hematoma at 12 hours from presentation (C, arrow).

Figure 2. Brain magnetic resonance imaging of hemorrhagic venous infarcts. Fluid-attenuated inversion recovery (FLAIR) sequence demonstrates bifrontal hematomas with confluent vasogenic edema (A, white box). T1-weighted postcontrast sequencing demonstrates diffuse cortical and meningeal enhancement with overlying vascularity (B, arrows). Echo-planar imaging (EPI) demonstrates additional punctate areas of susceptibility compatible with microhemorrhages (C, arrows).

(MRI) was negative for associated enhancement or microhemorrhages. Magnetic resonance angiography (MRA) of the brain showed nonspecific overlying vascularity. The patient was discharged to inpatient rehabilitation on hospital day 7. One month later, he presented with a 3-day history of intermittent confusion and 1-day history of lower extremity weakness. Blood pressure was elevated to 205/120 mm Hg, NIHSS score 0, and repeat noncontrast head CT showed progression of bifrontal hemorrhage with new subcentimeter hematomas in both frontal lobes (ICH score: 0, sICH score: 1). Repeat brain MRI demonstrated increasing macro- and new microhemorrhages with severe surrounding vasogenic edema, as well as new cortical and meningeal enhancement (Figure 2). Serological evaluations for infection and vasculitides were unremarkable. Cerebrospinal fluid analysis showed a modest elevation in protein (65 mg/dL) but was otherwise

unremarkable. The patient underwent anterior frontal lobe brain biopsy, which demonstrated leptomeningeal vasculopathy characterized by marked concentric obstructive intimal fibroplasia and associated with reactive cortical changes, including focal extensive cortical gliosis, rarefaction, and microfoci of cavitation. Though nonspecific, these findings were suggestive of secondary vascular congestion and ischemic injury, without evidence of a primary vasculopathy. Conventional cerebral angiography demonstrated a Cognard type IV dural arteriovenous fistula fed by bilateral ethmoidal arteries and draining directly into ectatic cortical veins (Figure 3). On retrospective review, there were some overlying vessels with intermediate flow characteristics, but the arteriovenous shunt was not clearly identified given the lower resolution of MRA. The fistula was successfully surgically obliterated via an uncomplicated left frontal craniotomy

Figure 3. Digital subtraction angiography of anterior cranial fossa dural arteriovenous fistula. Left internal carotid artery (ICA) injection, lateral view. Arterial phase imaging shows arteriovenous shunting with early filling of an ectatic venous channel (A, arrow, inset). Capillary phase imaging shows early cortical venous drainage (B, arrow). Venous phase imaging demonstrates venous drainage to an inferior anastomotic vein (C, arrow).

approach. There have been no complications for more than 8 months postprocedure. After rehabilitation therapy at a transitional care hospital, he was discharged home with assistance, with a modified Rankin Scale score of 3.

Discussion

This patient represents a unique case of a bilateral ethmoidal artery dAVF with cortical venous drainage, presenting with recurrent and progressive bifrontal intracerebral hemorrhages and additional stigmata of venous stasis. Due to their rarity, the literature regarding ACF-dAVFs consists mainly of case series. Features of this case consistent with those in the literature include male sex, elderly age, headache at symptom onset, arterial supply emanating from ophthalmic artery branches, leptomeningeal/cortical venous drainage, and the presence of variceal dilatation. However, many aspects of this case are reported in only a minority of cases: location in the ACF, bilateral arterial supply and venous drainage, hemorrhage as a presenting feature, resulting sinus thrombosis, and ultimately acceptable functional outcome following multifocal hemorrhages while anticoagulated.^{2,7} Furthermore, concurrent sinus thrombosis is found in only 1.5% of patients with dAVF based on a large Finnish case series.⁷

This case represents a rare presentation of a rare diagnosis and merits discussion of the wide differential diagnosis for progressive bifrontal cerebral hemorrhages. In elderly patients, low-impact trauma can produce hemorrhagic contusions in the inferior frontal and anterior temporal lobes, which are partly fixed to the anterior falx cerebri and greater sphenoid wings, as well as subdural hematomas, caused by shear injury to cortical bridging veins overlying patulous subarachnoid spaces. An underlying coagulopathy can further predispose to intracranial hemorrhage with minor trauma. In our patient, the presence of a right occipital scalp hematoma with left greater than right frontal hemorrhages highly supported the diagnosis of coup–contrecoup injury. However, subsequent imaging progression and evolution without additional trauma raised specific concern for an underlying

vascular abnormality. In particular, progressive macro- and microhemorrhages, symmetric bifrontal vasogenic edema, and cortical/meningeal enhancement are highly suggestive of progressive venous ischemia, which can be seen in the setting of high-flow vascular malformations and/or cerebral venous sinus thrombosis, both of which were ultimately identified in this patient. The deep bifrontal location of hemorrhage was not consistent with classic ruptured aneurysm (peripheral, subarachnoid) or hypertension (basal ganglia). Vasculopathies can present with similar but more diffuse imaging features. In this case, due to the localized bifrontal distribution and absence of systemic findings, an inciting vasculopathy was considered unlikely. Hemorrhagic conversion of underlying tumor, infection, or autoimmune disorders has also been described; however, our patient's initial imaging showed bland hematomas, and there was no underlying enhancement on follow-up studies to support an underlying parenchymal source. The biopsy showed nonspecific findings suggestive of arterialization of the venous system, as would be seen in a high-flow vascular malformation. Biopsy pathology showed the presence of venous ectasia and marked concentric obstructive intimal fibroplasia, which pointed toward venous hypertension, leading to further investigation with angiogram revealing the dural arteriovenous fistula.

Whether to perform catheter angiography can be a challenging decision given the inherent risks of the procedure. Secondary ICH score and the colloquially termed Hong Kong criteria are some tools that can help in making such a decision. A prospective Hong Kong-based study found that a history of hypertension and increasing age were independent negative risk factors for the detection of vascular anomalies via catheter angiogram, and this was most significant in cases of lobar hemorrhage.⁸ Of the patients with lobar ICH, 49% had positive angiography; however, angiographic yield was significantly lower in the older and hypertensive group (10%) compared with the younger and normotensive group (65%).⁸ The sICH score seeks to predict whether an ICH is secondary to an underlying vascular abnormality using a point-based

system. In the study defining the sICH score, a score of 1 conferred a 1.4% chance of vascular abnormality.⁹ Using tools such as these can be helpful, especially when data are limited and decisions have to be made quickly, yet both metrics appear to argue against use of catheter angiogram in our patient. However, a 10% yield is a nonzero probability, especially when diagnosis remains elusive and possible etiologies carry high morbidity. Zhu et al, in fact, advocate for the 9% yield in their hypertensive group as being sufficiently clinically significant to outweigh the procedural risk of angiography. This case highlights that such heuristics are supplements to, rather than substitutes for, clinical reasoning.

Understanding of the mechanisms of dAVF formation remains limited. Some propose these lesions are congenital and are unmasked by long-standing hemodynamic stress leading to rupture—a potential explanation for presentation in later life.¹⁰ Others suggest these are acquired lesions with aberrant angiogenesis incited by venous hypertension, trauma, surgery, chronic infection, and/or sinus thrombosis.² Also disputed is the etiopathogenesis of associated venous sinus thromboses. Primary thrombosis of cerebral sinuses may raise venous pressure causing arteriovenous shunts in the sinus walls. Alternatively, the abnormal pressure gradients in dAVF may alter cerebral blood flow, and secondarily, thrombose the sinus.^{11,12} In our case, the imaging and pathology findings suggest chronic venous hypertension with secondary development of superior sagittal sinus (SSS) thrombosis. In addition, the patient had a remote history of trauma related to falls 3 and 16 years prior and no history of prior neurosurgery or hypercoagulable state. In this setting, it is impossible to definitively know whether the AVF or SSS thrombosis came first.

It is important to recognize ACF-dAVF as a rare, but treatable, cause of ICH. Anterior cranial fossa dAVF should be considered in anyone presenting with cognitive decline, headache, seizure, and progressive intracranial hemorrhage, which in advanced stages can be associated with vasogenic edema and vascular thrombosis. Because magnetic resonance and CT angiography are less sensitive for detection of ACF-dAVF, conventional angiography should be strongly considered for detailed vascular assessment. Our case underscores the importance of maintaining a wide differential diagnosis in challenging clinical/imaging cases and shows that heuristics such as sICH score and the “Hong Kong criteria” are adjuncts to but not replacements for clinical reasoning. In conclusion, ACF-dAVF is a highly morbid yet treatable cause of recurrent intracranial hemorrhage, and surgical therapy has a very high rate of success and low risk of complications.²

Authors' Note

Written consent for publication of the patient's case and imaging was obtained from the patient.

Author Contributions

Stephen Johnson, Ben Cox, and Mai Lan Ho contributed to preparation and critical review of the manuscript. Meghan Murphy

contributed to critical review of the manuscript; Eugene Scharf contributed to critical review and supervision of the manuscript.

Declaration of Conflicting Interests

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