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Developmental Venous Anomaly Presenting with Headache, Transitory Symptoms and Non-Traumatic Calcarine Sulcus Subarachnoid Hemorrhage: A Case Report

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Abstract

Background: Developmental Venous Anomalies (DVAs) are the most common brain vascular abnormality. They are usually benign, incidental findings on brain images and asymptomatic.

Case: We report a case of a 48-year-old man with an acute headache and transient visual symptoms due to an occipital sulcus Subarachnoid Hemorrhage (SAH). Extensive diagnostic workup revealed an occipital DVA with an idiopathic mechanism of bleeding.

Conclusion: To the best of our knowledge this is the first reported case of an occipital sulcus SAH due to a DVA with an idiopathic mechanism of bleeding. The treatment of an intracranial hemorrhage due to a DVA will depend on the presenting symptoms and the pathophysiology of bleeding.

Keywords: CNS Venous angioma; Subarachnoid hemorrhage; Headache; DVA

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Introduction

Developmental Venous Anomalies (DVAs), previously known as venous angiomas, are the most common brain vascular abnormalities and are composed of a series of small radially arranged veins that drain into a dilated central vein [1,2] these are generally located in the frontal subcortical white matter and drain to the superficial or deep cerebral venous system [1,3]. Microscopically, the venous tissue is, for the most part, apparently normal with little changes of endothelial thickening and hyalinization [3,4].

Between 13% to 40% of DVAs are associated with Cerebral Cavernous Malformations (CCM), and a smaller percentage with other types of intracranial vascular malformations, dermatological syndromes or superficial head and neck venous malformations [5,6]. Isolated DVAs are generally incidental findings on brain images, asymptomatic and considered benign [7]. Nonetheless, case series and case reports have associated them with epilepsy, headache, focal neurological deficits and Intracranial Hemorrhage (ICH) [8-11]. A study of 422 patients with brain DVAs showed an annual risk of symptomatic bleeding from 0.34% to 1.28% [12]. Most ICHs associated with DVAs are intraparenchymal [7,12,13]. Cases of Subarachnoid Hemorrhage (SAH) in relation to DVAs have concomitant venous infarctions or intraparenchymal hemorrhage [13]. Lovrencic-Huzjan et al. [14] reported a case of basal cisterns SAH associated with a DVA. To the best of our knowledge there are no reports of cortical sulcus SAH associated with isolated DVA. Here we report an unusual case of an isolated occipital brain DVA presenting with cortical sulcus SAH.

Case Presentation

A 48-year-old Colombian male patient arrived at the emergency room with a history of a one day non-sudden, global, pulsating, headache of moderate intensity (7/10). One gram of acetaminophen was self-administered with short lasting relief. He complained of transient bilateral visual loss lasting several minutes at the beginning of the headache. He denied any history of recent or remote trauma, as well as previous headache or migraine diagnosis. The patient suffered from a stable chronic inflammatory demyelinating polyneuropathy in treatment with prednisolone 20 mg daily for the last 2 years. Upon admission his blood pressure was 125/84 mmHg, heart rate 72 bpm, respiratory rate 18 pm, afebrile and a general physical examination within normal limits. On neurological examination he was awake, orientated with normal cognitive function. His neuro-ophthalmologic examination was within normal limits, including visual acuity. Cranial nerves showed no deficit.

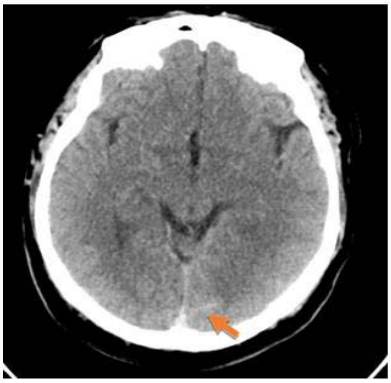

Figure 1: Non contrast brain computed tomography.
Axial cut showing a left occipital sulcal linear hyperdensity (red arrow).

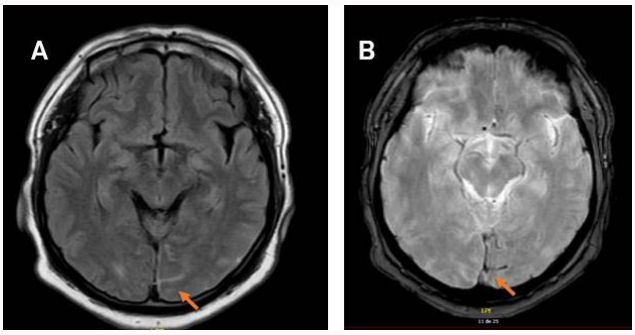

Figure 2: Simple brain MRI.
A: MRI FLAIR sequence showing a left occipital sulcal linear hyperintensity (Red arrow). B: MRI GRE sequence showing a left occipital linear hypointensity (Red arrow).

On motor examination, his strength was 4/5 in all four limbs (distal and proximal) with generalized hyporeflexia and a bilateral neutral plantar response. His sensory examination revealed no abnormalities and cerebellar signs were absent. There was no evidence of meningeal signs or neck stiffness.

A non-contrast brain Computed Tomography (CT) (Siemens 128-channel CT) was performed showing a linear hyperdensity in the left calcarine sulcus (Figure 1). Laboratory work up including platelets, prothrombin time, and activated partial thromboplastin time were within normal limits. A nasopharyngeal swap smear for SARS-CoV-2 virus was negative. Further work up with a contrast enhanced brain magnetic resonance imaging (MRI Phillips 1.5 Tesla) showed hyperintensity on FLAIR (Fluid-attenuated inversion recovery) sequences and hypointensity on GRE (Gradient echo) sequences in the left calcarine sulcus with no overlying images of CCM or Arterio-Venous Malformation (AVM) (Figure 2A,B). Contrast enhanced arterial and venous brain MRI angiography (Phillips 1.5 tesla) showed hypoplasia of the left anterior cerebral artery and a fetal origin of the right posterior cerebral artery without evidence of an arterial-venous malformation or fistula (Image 3A). The venous phase revealed hypoplasia of the right lateral sinus and an image compatible with a DVA in the left subcortical occipital region. There was no evidence of thrombosis or stenosis of the brain's venous system (Figure 3B and 3C).

Findings of cerebrospinal fluid on lumbar puncture were: red blood cells 1440 mm^3 (100% fresh), proteins 114.6 mg %, 0 leukocytes

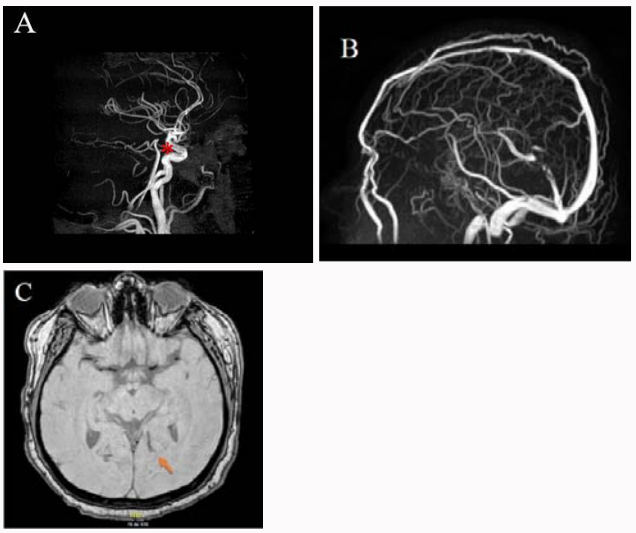

Figure 3: Contrast-enhanced brain MRA arterial phase (A) and venous phase (B and C).
A: Contrast enhanced MRA arterial phase showing A1 segment hypoplasia of the left ACA and a fetal origin of the right posterior cerebral artery (*). Note the absence of vascular malformations.
B: MRA venous phase showing right lateral sinus hypoplasia.
C: MR showing dilated central vein of the DVA in the left occipital lobe (Red arrow).

per mm^3, non-reactive VDRL (Venereal Disease Research Laboratory) and negative stains. Oral acetaminophen 1 gr TD was initiated with headache resolution within 48 h. On day 5 after admission, an arterial and venous brain conventional angiography was performed at another institution showing the same results as in the initial MRIs (Images not available). The patient remained without headache or visual symptoms and was discharged home with acetaminophen 1 g orally in case of headache, prednisolone 20 mg 8 am for his underlying disease and neurological outpatient follow up. The patient was lost to follow-up, but there was no record of new admissions to the emergency room in our institution.

Discussion

The patient presented a headache syndrome with red flags: acute new onset headache with neurologic symptoms. Initial neuroimaging studies showed left occipital calcarine sulcus SAH. Further work up led to the conclusion of a secondary headache syndrome: An occipital sulcus SAH due to an isolated DVA. Sudden or acute severe headache and transient focal symptoms are not unusual in sulcus or convexity SAH [15,16]. What is unusual in this case is its cause, the location of the DVA and the presumed pathophysiological mechanism of bleeding.

In a study of 23 patients in Argentina, the clinical manifestations of convexity SAH due to diverse etiologies included headache (60%), sensory and/or motor symptoms (47%), and epileptic seizures (30%). Six (26%) patients had thunderclap headache. Khurram et al. [17] described a series of 41 patients with convexity SAH transient neurological symptoms in 63% and the etiologies included: Cerebral Amyloid Angiopathy (CAA) (39%), Reversible Cerebral Vasoconstriction Syndrome (RCVS) (17%), cerebral venous thrombosis (10%), intracranial atherosclerotic disease (10%) and posterior reversible leukoencephalopathy syndrome (5%). The cause was not identified in 20% of the cases. Kumar et al. [18] reported in

29 patients with convexity SAH abrupt onset severe headache as the main manifestation in patients <60 years of age and in relation to SCVR. In patients >60 years of age the main cause was CAA with transient sensory and motor symptoms [19].

DVAs are usually asymptomatic but have been associated with ICH. The vast majority of ICH are intraparenchymal and the few SAH reported in the literature have been associated with venous infarctions, intraparenchymal hemorrhages, other brain vascular abnormalities and one case of basal cisterns SAH [1,3,4,7-9,14]. Although DVAs are almost exclusively supratentorial, their most frequent location is the frontal lobe, only a few of them have been reported in the occipital lobe [1,3].

There are three described pathophysiological mechanism which could explain bleeding of a DVA: Imbalances of blood inflow/outflow, venous thrombosis/stenosis and cryptogenic/idiopathic. The latter being the less commonly reported [20]. An imbalance with increased blood inflow and/or decreased outflow in the DVA can cause increased pressure and rupture [20-22]. The increased inflow maybe due to an Arteriovenous Malformation (AVM) that drains into the DVA or micro shunts within it [20-23]. The decreased outflow can be due to a remote increase of pressure in the brain's venous system, as it occurs in AVMs or arteriovenous fistulas [20,24]. Obstruction of the venous channel of the anomaly, as in cases of thrombosis or stenosis of the DVA or the cerebral vein into which it drains, can cause increase venous pressure or rupture [20-27]. Lastly, a small percentage of DVAs bleed and extensive neuroimaging work up do not reveal any additional abnormality [20-27].

Our patient underwent extensive neuroimaging diagnostic work up. A non-contrast CT, contrast enhanced MRI, contrast enhanced brain arterial/venous MRI and brain arterial/venous conventional digital substraction angiography showed only an occipital DVA with an overlying sulcus SAH. The final diagnosis made was a secondary headache syndrome due to a sulcus SAH with an isolated symptomatic occipital DVA with an idiopathic bleeding mechanism.

The management of ICH or SAH associated with a DVA depends on the symptoms, size, location, and pathophysiological mechanism of the bleeding [20,28]. As the patient presented with occipital sulcus SAH, with mild symptoms that rapidly resolved, with an idiopathic bleeding mechanism, a low probability of recurrence was entertained. Symptomatic management with oral simple analgesics and outpatient follow-up was prescribed.

Conclusion

To the best of our knowledge this is the first reported case of a sulcus SAH due to an isolated occipital DVA with an idiopathic mechanism of bleeding. Sulcus/convexity SAH is in the differential diagnosis of sudden or severe headache with transient neurological symptoms. DVAs could be a rare cause of sulcus or convexity SAH. Finally, in the presence of a symptomatic DVA with ICH, it is important to understand the bleeding mechanism to guide treatment decisions.

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