Open Journal of Clinical & Medical Images

www.jclinmedimages.org

Case Report

Open Access, Volume 3

Developmental venous malformation: A "Do-Not-Touch" lesion

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Received: Jun 10, 2023

Accepted: Jul 13, 2023

Published: Jul 20, 2023

Archived: www.jclinmedimages.org

Abstract

The most common slow-flow venous malformation in the brain are Developmental Venous Anomalies (also known as DVAs). The vast majority of DVAs are harmless, while seldom they can become symptomatic, leading to a variety of pathologies. DVAs can vary greatly in size, location, and angio-architecture. Imaging evaluation is the best tool in evaluating these types of lesions when they become symptomatic.

Keywords: Developmental; Brain; Vascular; Malformation.

Case presentation

We present the case of a 60-year-old patient who comes to the emergency department complaining of dizziness. Non-contrast head CT examination reveals a linear, spontaneously hyperdense image located in the right precentral gyrus, extending from the cortical level to the deep white matter, with associated perilesional edema (Figure 1); the presumptive diagnosis is venous thrombosis or anomaly of venous development.

Admission on Neurology department and further imaging investigations with MRI with dedicated protocol were recommended.

MRI examination with T1 sequences with contrast (Figure 2A and 2B), FLAIR (Figure 3), SWAN (Figure 4), DWI/ADC (Figure 5), showing a straight posterior frontal dilated venous tract with

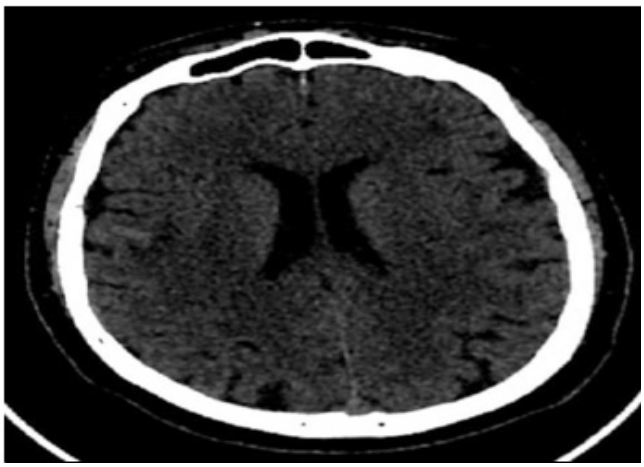

Figure 1: Non- cotract axial head CT examination reveals a linear, spontaneously hyperdense image located in the right precentral gyrus, extenfding from the cortical level to the deep white matter, with associated perilesional edema.

"caput-medusae'' head branching, extended to the deep white matter level, which spills into a cortical vein draining into the superior sagittal sinus with adjacent changes in hyperintense FLAIR signal; no filling defects on post-contrast sequences, no diffusion restriction; imaging appearance suggesting a venous developmental anomaly with adjacent gliotic changes.

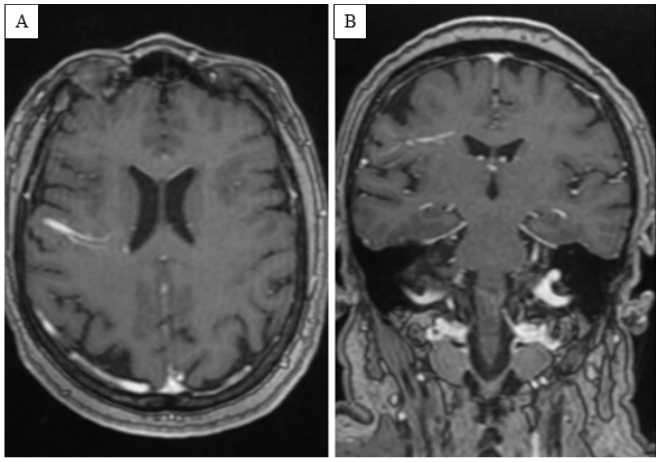

Figure 2: Axial **(A)** and coronal **(B)** T1 post-contrast MRI sequences venous phase showing straight posterior front dilated venous tract head branching, extended to the deep white matter level, which spills into a cortical vein draining into the superior sagittal sinus.

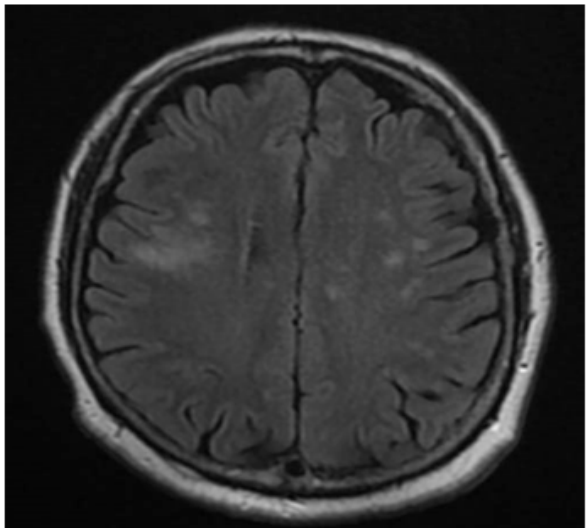

Figure 3: Axial MRI FLAIR sequence showing adjacent gliotic changes in hyperintense signal.

Discussion

A Developmental Venous Anomaly (DVA) is an extreme variation of a transmedullary vein composed of a radial complex of medullary veins resembling a "Medusa head" appearance, which converges into a "collector" vein that eventually drains into either the superficial or deep or cerebral venous system; superficial type drains into the cortical veins and dural venous sinuses, while deep type DVA primarily enters the Galenic system [1,2]. It is described as being the most common type of slow-flow venous malformation in the brain, with an estimated incidence between 2% and 6,4%, and the vast majority of them are asymptomatic [1].

Histologically they are composed of dilated venous channels

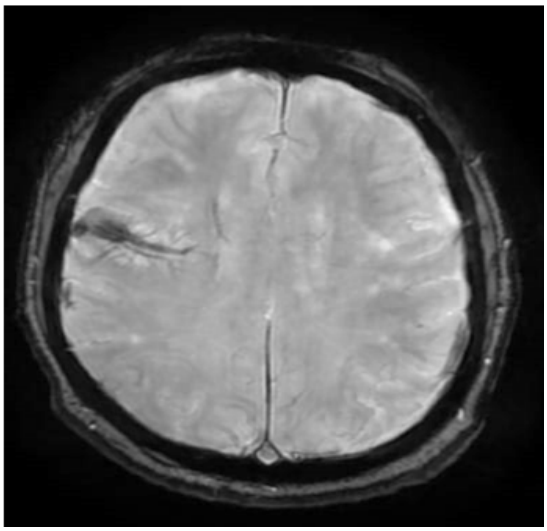

Figure 4: Axial MRI SWAN sequence showing a straight posterior frontal dilated venous tract with "caput-medusae" head branching, extended to the deep white matter level, which spills into a cortical vein draining into the superior sagittal sinus.

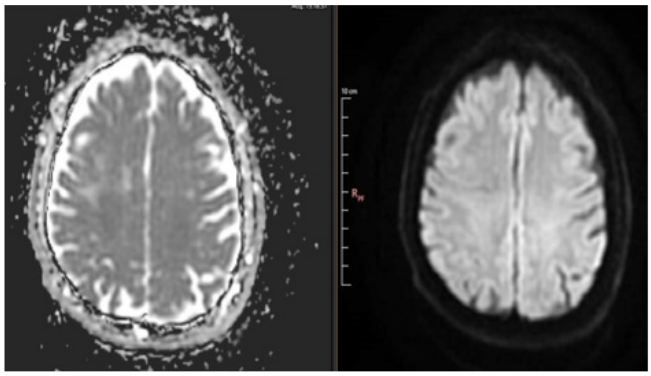

Figure 5: MRI DWI/ADC sequences showing no diffusion restriction.

interspersed in the white matter, with simple or complex variations in venous architecture and drainage patterns [1]. Over time, exposure to higher venous pressure may cause vascular problems and patients may become symptomatic [1]. They can be supratentorial, affecting the cerebral cortex, or infratentorial, affecting the brainstem or cerebellum; however, the majority of them are supratentorial lesions [3]. When these lesions become symptomatic, they can lead to seizures, hemorrhage, headache, and focal neurological deficits [3]. Studies suggest that supratentorial lesions are more commonly associated with seizures, whereas infratentorial malformations are associated with focal neurological deficits [3]. Prior to the development of Computed Tomography (CT) and Magnetic Resonance Imaging (MRI), DVAs were thought to be uncommon causes of intracranial hemorrhage and seizures [4]. Garner et al. reported that the risk of true DVA-related hemorrhage is 0.22% per year [4]. DVAs are associated with cavernous malformation in up to 13-40% of cases; these are thought to be responsible for the vast majority of hemorrhagic cases [4].

As a general rule, it is not recommended to treat angiomas located near the venous drainage of the brain [5]. If a patient has associated cavernous malformations that require surgery,

the angioma should not be touched [5]. Surgery for the angioma itself is only advised if there is evidence of bleeding or if the patient is experiencing uncontrollable seizures that can be directly linked to the lesion [5].

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