

# Spontaneous thrombosis of developmental venous anomaly (DVA) with venous infarct and acute cerebellar ataxia

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**Abstract** Developmental venous anomaly (DVA), formally known as venous angioma, is a congenital anatomic variant of the venous drainage of the brain. Although they typically have a benign clinical course and a low symptomatic rate, thrombosis of a drainage vein may occur, leading to potentially debilitating complications. We report a unique case of spontaneous thrombosis of a posterior fossa developmental venous anomaly with cerebellar infarct in a 61-year-old man who presented with acute onset cerebellar ataxia. DVA thrombosis was well-depicted on CT and MR studies. Patient was put on anticoagulant therapy and complete recanalization was seen on follow-up imaging.

**Keywords** Developmental venous anomaly · Thrombosis · Vascular

## Introduction

Developmental venous anomaly (DVA) is fairly common and generally benign congenital vascular abnormalities, with a

reported incidence between 0.7 and 2.6 % in the general population [1, 2]. DVAs represent a purely venous entity, an anomalous venous disposition due to the absence of normal pial or subependymal veins. There is still controversy as to how DVAs develop, though it is generally accepted that they form during intrauterine life [3]. DVAs have a characteristic appearance on CT and MR studies, and angiography is seldom needed. Digital subtraction angiography (DSA) is generally reserved for cases presenting with ischemic or hemorrhagic infarction, or whenever an associated vascular malformation is suspected on CT or MRI. Unlike other congenital malformations, DVAs rarely produce symptoms; therefore, in most cases, they are diagnosed incidentally. Nonetheless, they rarely cause symptoms due to either mechanical compression to adjacent intracranial structures [4, 5]. Thrombosis of a DVA is extremely rare, and most symptomatic complications arise from DVAs that are arterialized or associated with other vascular malformations, like cavernous malformation or arteriovenous malformations. In this paper, we report the imaging findings in a case of DVA with thrombosis of the draining vein in the left cerebellar hemisphere and resultant venous infarct.

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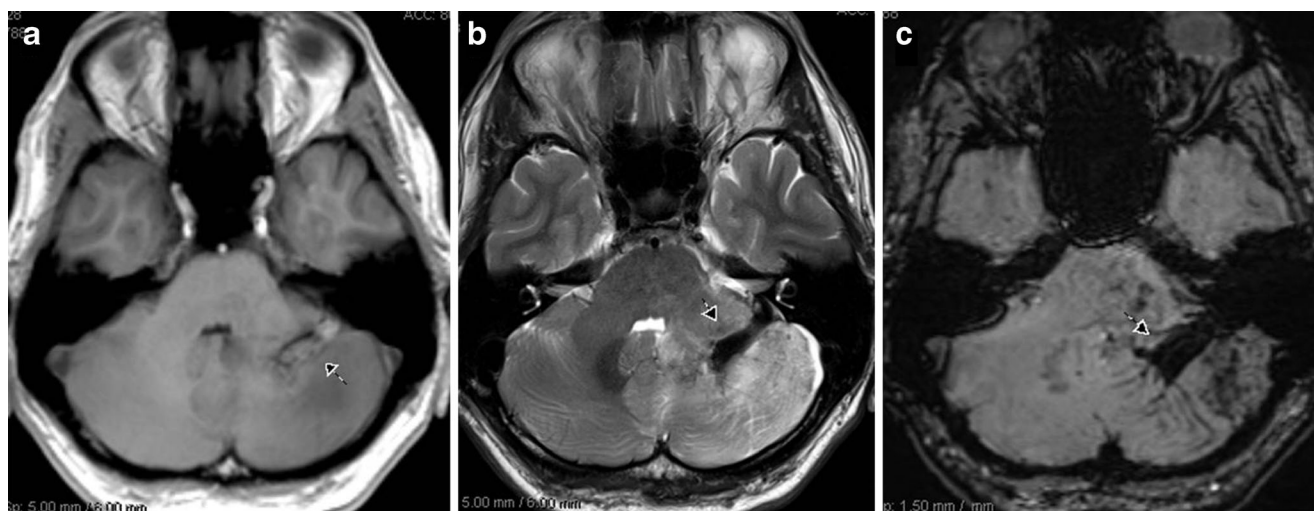
## Case report

A 61-year-old man presented to an emergency room with acute onset cerebellar ataxia and syncope. He had experienced headaches, progressive dizziness, and nausea for few days. He denied previous seizure and had no cardiovascular risk factors. Laboratory values, including coagulation tests, were within normal limits. Cranial computed tomography (CT) scan revealed a large tubular hyperdense structure in the left cerebellar hemisphere converging toward the left sigmoid sinus (Fig 1). Additionally, mild left-sided cerebellar edema was present. Magnetic resonance imaging (MRI) performed



**Fig. 1** Axial unenhanced head CT scan through the posterior fossa shows a hyperdense tubular structure in the left cerebellum (*arrow*). Additionally, there is mild left-sided cerebellar edema

subsequently revealed an intrinsic T1-hyperintense tubular structure in the left cerebellum (Fig 2a) with corresponding dark signal on T2-weighted images (Fig 2b), consistent with intraluminal thrombus (intracellular methemoglobin). Signal drop-out with blooming was seen on the susceptibility-weighted images (SWI; Fig 2c). There was extensive surrounding vasogenic edema and mass effect with no restricted diffusion, consistent with venous infarct. Gadolinium-enhanced T1-weighted images showed the nonenhancing



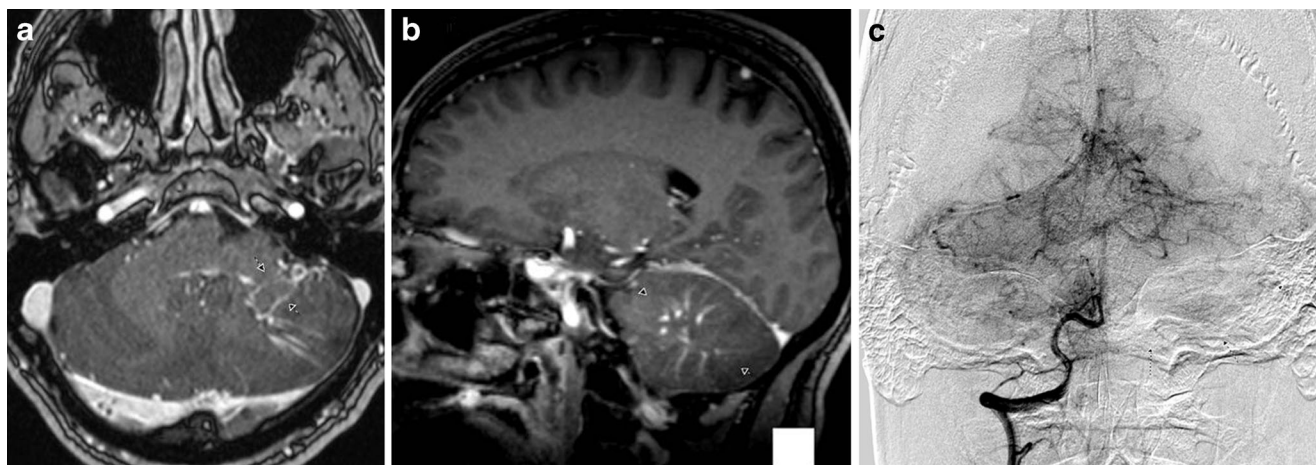
**Fig. 2** Three axial MR images through the posterior fossa. Unenhanced T1-weighted image reveals an intrinsic T1-hyperintense tubular structure in the left cerebellum (Fig 2a; *arrows*) with corresponding dark signal on T2-weighted images (Fig 2b; *arrows*), consistent with intraluminal thrombus (intracellular methemoglobin). Signal drop-out with blooming

thrombosed draining vein along with multiple enhancing medullary veins, with caput medusa appearance, confirming the diagnosis of DVA (Fig 3a, b). Four-vessel cerebral angiography during the venous phase demonstrated the lack of opacification in the territory of the thrombosed DVA (Fig 3c). The patient's neurological status improved gradually with anticoagulation therapy over a period of 8 days with residual mild left-sided ataxia, and he was discharged on coumadin and antihypertensive medications. Follow-up MRI after 3 months showed recanalization of the thrombosed vein (Fig 4a, b) and resolution of cerebellar edema. No underlying cavernous malformation or AVM was identified on catheter angiogram or follow-up MR imaging.

## Discussion

Since the introduction of magnetic resonance (MR) imaging, DVAs of the brain have become the most frequently detected cerebral vascular malformation [6]. DVA represents a dilatation of a transcerebral vein that acts as a collector for small venules; it is thought to develop in response to the absence of one of the usual venous drainage routes [7]. The appearance of a DVA on angiography is synchronous with the normal venous phase. It drains normal brain and the underdevelopment of the usual pattern is clearly visible in most cases. A developmental venous anomaly may drain a cortical area toward the deep venous system or a deep area toward the superficial venous system [7, 8]. DVAs are associated with sporadic cavernous malformations (CM) in 13 to 40 % of cases and are encountered both in the pediatric and adult populations [3]. They have a characteristic appearance on

is seen on the susceptibility-weighted images (SWI; Fig 2c). Note the multiple linear vessels with signal drop-out converging towards the large draining vein on the SW image, representing the *caput medusa*. Surrounding parenchymal edema and mass effect is best appreciated on the T2-weighted axial images



**Fig. 3** Gadolinium-enhanced T1-weighted images shows the nonenhancing thrombosed draining vein (Fig 3a; *arrows*) along with multiple enhancing medullary veins, with caput medusae appearance (Fig 3b; *arrows*),

confirming the diagnosis of thrombosed DVA. Catheter angiogram image during the venous phase (Fig 3c; *arrows*) shows lack of opacification in the territory of the thrombosed DVA, in the left cerebellum

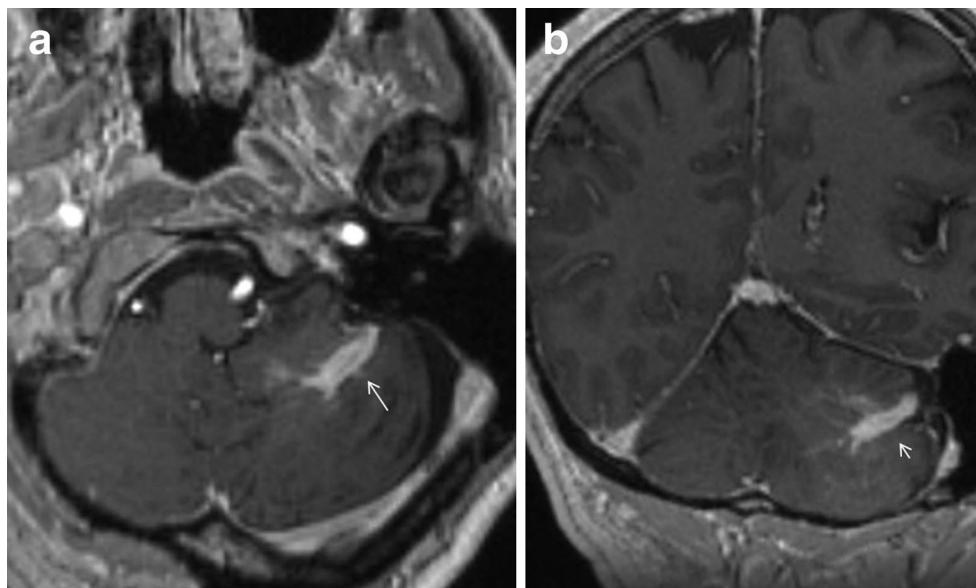
CT and MR studies, and angiography is seldom needed. Digital subtraction angiography (DSA) is generally reserved for cases presenting with ischemic or hemorrhagic infarction, or whenever an associated vascular malformation is suspected on CT or MRI.

Regardless of the imaging modality employed, the diagnosis of a DVA relies on demonstrating a typical caput medusae draining into a collecting vein. Because of its higher temporal resolution, DSA remains the best imaging modality to study the hemodynamic behavior of DVAs. On noncontrast CT, the collecting vein of the DVA may appear to be isodense or slightly hyperdense to the cortex if patent, or markedly hyperdense if acutely thrombosed. In addition, noncontrast CT may disclose an associated hemorrhage, parenchymal calcifications, and atrophy or white matter lesions in the drainage territory of the DVA [9]. Both the collecting vein

and the caput medusae enhance after the administration of contrast material and are best demonstrated by thin-section CT venography. Noncontrast T2- and T1-weighted MRI may demonstrate flow voids and phase-shift artifact produced by the collecting vein of a DVA and by the larger venous radicles of the caput medusae. As with CT, the caput medusae and the collecting vein readily enhance after gadolinium administration on T1-weighted sequences. Because CMs are frequently associated with DVAs, they should be specifically sought for using hemosiderin or deoxyhemoglobin-sensitive sequences such as SWI. CMs may be difficult to identify in the setting of a DVA presenting with acute hemorrhage and may only become apparent after the hemorrhage is fully reabsorbed, typically 3 to 6 months after the initial event [3].

Unlike arteriovenous malformations and to a lesser extent cavernous malformations, DVAs rarely produce symptoms

**Fig. 4** Post-contrast images from a 3-month follow-up MRI scan shows the recanalized enhancing draining vein (Fig 4a, b; *arrows*) along with multiple enhancing medullary veins of the developmental venous anomaly



and in most cases are therefore diagnosed by chance [6]. Cavernous malformations (CM) are now thought to be responsible for the vast majority of symptomatic cases previously attributed to DVAs themselves. Hemorrhagic or ischemic infarction around a DVA may result from acute thrombosis of the collecting vein, though such complications are very rare. A review of the literature revealed 19 documented cases of symptomatic thrombosed DVAs presenting with venous ischemic infarction (53 %), parenchymal hemorrhage (37 %), and subarachnoid and intraventricular hemorrhage (5 %) [3]. Management is usually conservative with anticoagulation therapy, with decompression surgery for evacuation required in few complicated cases. Clinical outcome is favorable in the majority of patients with complete recovery or persistence of mild neurological symptoms. Although no large series or controlled studies exist to support systemic anticoagulation over conservative management in cases of thrombosed DVAs, experience gained from treatment of cortical or dural venous sinus thrombosis suggests that systemic and prolonged anticoagulation should also be of benefit in preventing clot propagation, in favoring recanalization, and in promoting reversibility of the symptoms in such cases. Finally, screening for prothrombotic conditions should be carried out as with cortical and dural venous sinus thrombosis [10, 11]

In conclusion, DVAs can occasionally give rise to significant symptoms due to thrombosis of the draining vein. We evaluated comprehensive imaging findings in a challenging case of DVA associated with nonhemorrhagic infarction. The familiarity with imaging findings of complicated DVAs is essential for accurate diagnosis and appropriate management.

**Conflict of interest** The authors declare that they have no conflict of interest.

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