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CORRESPONDENCE

Isolated Developmental Venous Anomaly of the Pons with Transpontine Drainage: Case Report

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Introduction

Initially known as venous angioma, Developmental Venous Anomaly (DVA) is a persistent variant of fetal brain venous drainage [1, 2]. Unlike cerebellar DVA with or without brainstem extension, isolated pontine DVA is a rare condition [3, 4]; and as per our knowledge, its presentation with exclusive transpontine drainage has never been described before. The authors present the case of an isolated pontine DVA with a selective transpontine drainage discovered incidentally; the clinical presentation, the radiological findings, and the management are discussed.

Case Report

A 77-year-old woman presented to the emergency unit with a transient constellation of nonspecific symptoms, and no clearly defined neurological symptoms or signs. The brain CT scan and MRI sequences: T1 turbo spin echo (TSE) with and without injection of gadolinium (Gd), T2 TSE, T2 gradient echo as well as MRI diffusion images excluded an ischemic or hemorrhagic lesion of the brain, but revealed an incidental pontine vascular lesion. CT scan of the brain (Fig. 1a, b) revealed a diffuse enhancement of the center and right side of the pons; with an adjacent vascular-like enhancement located in the right pontine tegmentum. Sagittal T1 TSE (Fig. 2a) revealed a hypointense lesion in the pontine tegmentum; this latter showed a vascular-like enhancement (Fig. 2b). Axial T1 TSE Gd images showed a vascular-like enhancement presenting radicular divisions in the deep pontine tegmentum having a caput medusae appearance (Fig. 3a), without extension to the cerebellum or the fourth ventricle. This vascular enhancement passes through the right pontine tegmentum, and exits in the right pontocerebellar cistern (Fig. 3b). Axial T2 TSE images (Fig. 4a, b) showed a linear hypointense signal following the pathway of the right pontocerebellar cistern, the right pontine tegmentum, and directing towards the anterior part of the fourth ventricle's floor (*arrows* in Fig. 4a, b). Gradient echo T2 images (Fig. 5) clearly showed a hypointense vascular-like signal in the right pontine tegmentum, with deep hypointense radicular divisions. The radiologic presentation was consistent with an isolated DVA of the pons with a transpontine drainage, without associated bleeding or stenosis of the main collector vein. The laboratory exams revealed a moderate hyponatremia, which was corrected with intravenous fluids. A conservative management was adopted with complete relief of symptoms. The patient was discharged two days after admission.

Discussion

DVA is a persistent variant of fetal brain venous drainage [1, 2]. This is thought to be due to an arrest of formation or thrombosis of the developing venous drainage system,

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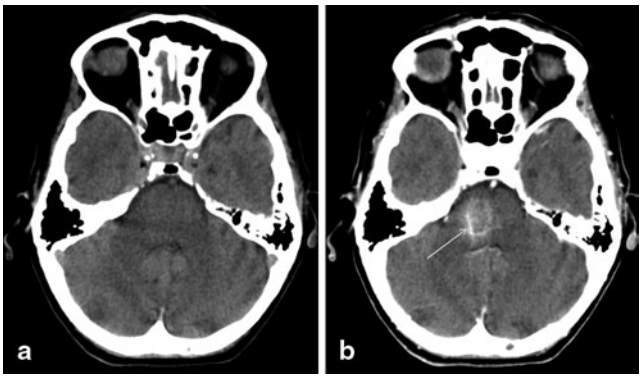

Fig. 1 **a** Axial noncontrast brain CT scan. **b** Axial contrast brain CT scan. There is a vascular-like enhancement located in the right pontine tegmentum, extending toward the floor of the fourth ventricle (*arrow* in **b**); this represents the DVA drainage. Around this vascular-like enhancement, and mostly at its left, there is a diffuse contrast enhancement without enlargement of the pons, nor mass effect on the peripheral structures; representing a site of hyperemia due to the terminal divisions of the DVA

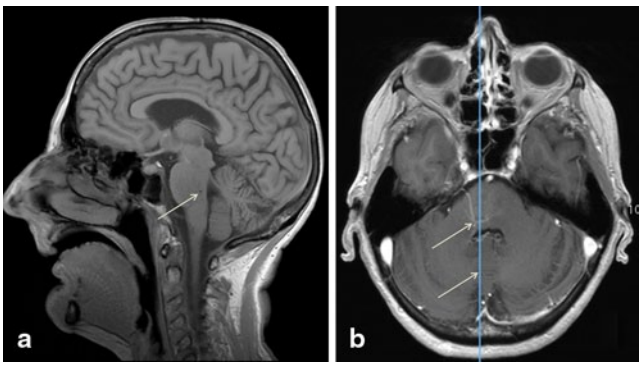

Fig. 2 Sagittal T1 TSE image (**a**) and Axial T1 TSE Gd image (**b**). The reference line in blue (*lower arrow* in figure **b**) is passing through drainage vein (*upper arrow* in figure **b**). The drainage vein appears as a hypointense structure on noncontrast images (**a**), and shows a vascular-like contrast enhancement with deep terminal division branches (**b**)

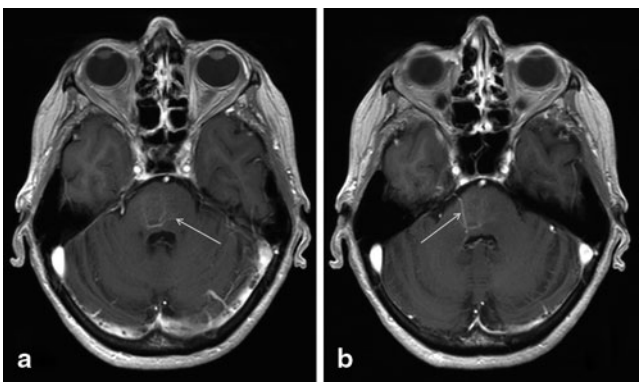

Fig. 3 Axial T1 TSE Gd images (**a** and **b**). This illustrates a vascular-like enhancement, with deep radicular divisions in the pontine tegmentum (*arrow* in **a**) corresponding to the radicular branches of the DVA. In **b** (*arrow*), we can clearly identify a vascular enhancement, passing through the right pontine tegmentum, and exiting the pons at the level or the right pontocerebellar cistern. This represents the drainage vein of the DVA. There is no extension toward the cerebellar regions or to the fourth ventricle

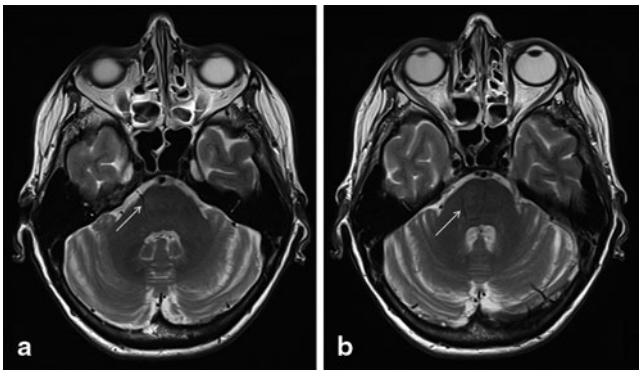

Fig. 4 Axial T2 TSE images (**a** and **b**). This shows a linear hypointense structure exiting the pons through the right ponto-cerebellar angle, and located essentially in the right pontine tegmentum, corresponding to the drainage vein of the DVA (*arrows*). There is no extension towards the cerebellar regions or to the fourth ventricle

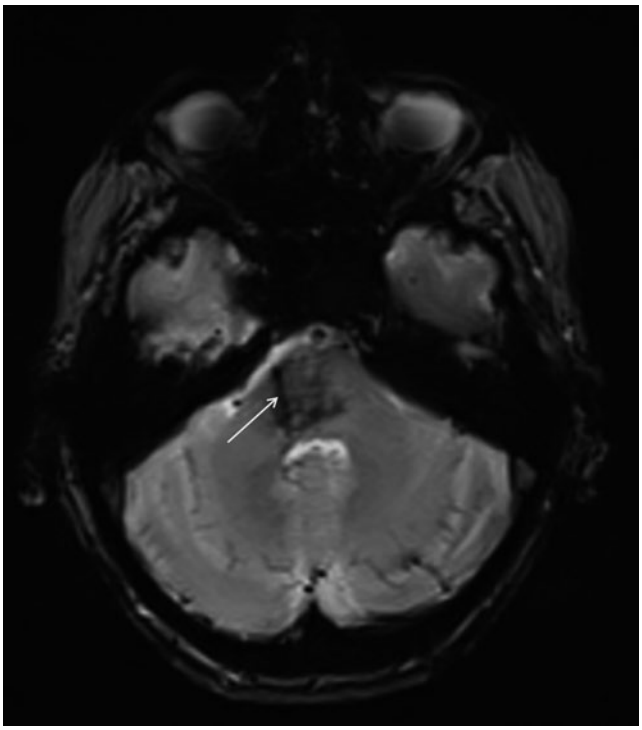

Fig. 5 Axial gradient echo T2 image (T2*). This illustrates a linear shape hypointensity passing through the right pontine tegmentum, with an exit point in the right ponto-cerebellar cistern. This corresponds to the DVA collecting vein. This latter shows hypointense divisions in the deep pontine tegmentum representing the radicular branches of the DVA. There is no diffuse hypointensity, which excludes an associated telangiectasia

which leads to a secondary dilatation of peripheral venules, with classic aspect of caput medusae [5–7]. DVA is the most frequent cerebrovascular malformation [8]. Its incidence varies between 0.48 % [9, 10], up to 4.05 % in some autopsy series [11]. DVA can appear as an isolated finding or can be in association with other malformations such as cavernous angioma [12–21] or capillary telangiectasia

[22, 23]. Among nonarterial vascular malformations, the most frequent association has been described between DVA and cavernous malformations [24, 25]. Recently, some authors have reported cases or malformations appearing in a triad composed of capillary telangiectasia, cavernous angioma, and DVA [5, 25, 26]. In our report, the DVA was observed as an isolated vascular anomaly. This anomaly was located within the pons, with an exclusive transpontine drainage (Fig. 3a, b). According to the literature, in one-third of cases, DVA are located in the posterior fossa [10, 27, 28], predominantly in the cerebellum with a prevalence of 14–36% [4, 29]. Around 3–8.7% of DVA [4, 29] are located in the brainstem; only a 1.5% prevalence is reported of pontine location [29]. As per our knowledge, a case of isolated exclusive pontine DVA with selective transpontine drainage has never been reported. Four reported cases of exclusive pontine DVA with selective transpontine drainage were associated with other vascular malformations [24, 25, 30, 31]. There is one reported case of isolated exclusive pontine DVA, with collector vein draining through the fourth ventricle, into the vein of Galen [32].

The radiological findings of DVA are well established [1, 9]. Independent of the imaging modality, the diagnosis relies on demonstrating a typical caput medusae draining into a collecting vein. The gold standard method remains the digital subtraction angiography, but in most cases CT and MRI allow the diagnosis of such lesions; the last one being superior to demonstrate associated parenchymal abnormalities. Noncontrast CT reveals the collecting vein as isodense or slightly hyperdense (if patent) or markedly hyperdense (if thrombosed). Noncontrast T2 and T1 weighted MRI may reveal flow voids and phase-shift artifact due to the collecting veins and the larger venous radicles of the caput medusae. After contrast administration, the caput medusae and the collecting veins enhance both on CT and T1 weighted MRI. In case of a pontine location, as in our report, capillary telangiectasia may be evoked [35–37]. It may be difficult to differentiate between the two lesions. It should specifically sought for using hemosiderin or deoxyhemoglobin sensitive sequences such as gradient echo T2 weighted images or susceptibility weighted images (SWI) [38]. The radiological features of telangiectasias on MRI imaging are hyperintensity on T2 SE (or T2 TSE), which can often be absent as well [36, 37]. Diffuse hypointensity on gradient echo T2 weighted images, on the other hand, is more specific [36]. In our report, these two specific MRI observations for capillary telangiectasia were absent (Figs. 4a, b, and 5). Moreover, the different MRI sequences showed a large pontine drainage vein (Figs. 3b, 4b, and 5), with radicular branches in the deep pontine tegmentum (Figs. 3a and 5). This remains consistent with a DVA of the pons.

DVA classically remains asymptomatic, and is frequently encountered as an incidental radiological finding [8] on cerebral images performed for unrelated symptomatology. The clinical significance of DVA and its management remain controversial [8, 33]. The most frequent related symptoms reported in hospital-based clinical studies are headaches (50.8%), focal neurologic deficits (39.7%), and seizures (30.2%) [34]. Given the benign nature of DVA, their causality effect toward these symptoms is uncertain. Garner et al. [4], in their studies on 100 patients with DVA, described the possible causality relation of neurological symptoms to a DVA: headaches in 4/36, seizures in 5/23, and focal neurological deficit in 8/41. The hemorrhagic risk of DVA is negligible [12]. It varies from 0.22 to 0.68% per year [12, 29]. Following recent studies, supratentorial and posterior fossa DVA carry the same hemorrhagic risk [29, 34]. In hemorrhagic cases, the DVA is usually associated with a related mature or immature cavernous malformation, often found in the venous angioma's radicular branches, responsible for the bleeding [25]. The hypothesis for bleeding in isolated cases of DVA is possibly due to stenosis or thrombosis of the collector vein [8, 25] with consecutive intracranial venous hypertension within the radicular veins of the DVA leading to microbleeds.

In our report, there was neither infarct nor bleeding in the brainstem. Our patient presented with a transient constellation of symptoms that could not be ascribed to the pontine location of this DVA. We effectively considered this diagnostic as an incidental finding.

DVA are known to be anatomic variations draining normal brain (or brainstem) parenchyma [2, 5]. Hence, the surgical resection of these lesions or radiosurgery on DVA is contraindicated [39]. Resection of DVA carries the risk of hemorrhage, venous infarction, or extensive edema of normal brain, brainstem, or cerebellar tissue [28, 40, 41]. The only possible indications for surgery in cases of DVA are the rare instances when they present with an extensive intraparenchymal hematoma with mass effect [42–44].

Conclusion

The authors describe the first reported case of isolated pontine DVA with exclusively transpontine drainage, discovered incidentally. In the absence of an ischemic or hemorrhagic presentation, the clinical significance of DVA remains uncertain. Hence no surgical treatment is warranted for these vascular lesions.

Conflict of Interest The authors attest that there is no conflict of interest for the submission and publication of the present case report.

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