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Case report

Unusual location of developmental venous anomaly within fourth ventricle causing obstructive hydrocephalus – A case report

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1. Introduction

Most developmental venous anomalies (DVA) are asymptomatic and found incidentally during cranial imaging exams. However, though quite rare, symptomatic DVAs with manifestation of parenchymal hemorrhage, hydrocephalus, facial spasm, trigeminal neuralgia and venous congestion show its clinical significance and should not be ignored. Herein, we presented a case with obstructive hydrocephalus owing to fourth ventricle outlet obstruction by DVA and recovered well after cerebrospinal fluid (CSF) diversion.

2. Methods

A 28-year-old female from Vietnam with a history of chronic headache came to our clinic owing to worsening of the headache and unsteady gait for 6 months. Physical examination showed papilloedema with unremarkable lab tests. Brain magnetic resonance imaging (MRI) series showed obstructive hydrocephalus with dilated lateral ventricles, third ventricle and fourth ventricle and a DVA over right cerebellar hemisphere (Fig. 1). Magnetic resonance venography (MRV) showed tortuous vascular lesion occupying the outlet of fourth ventricle, so we arranged brain digital subtracting angiography (DSA) to exclude the possibility of cavernous malformation (CM) and arteriovenous malformation (AVM). The brain DSA showed right cerebellar DVA with typical caput medusae appearance and no other related vascular lesion (Fig. 2). Engorged transmedullary veins impeded the outlet of fourth ventricle, causing subsequent obstructive hydrocephalus. After thorough examination and pre-operative preparation, ventriculo-peritoneal shunt performed 2 days later. The patient recovered well with improved headache and walked steadily without further discomfort and received regular neurosurgical clinic visit.

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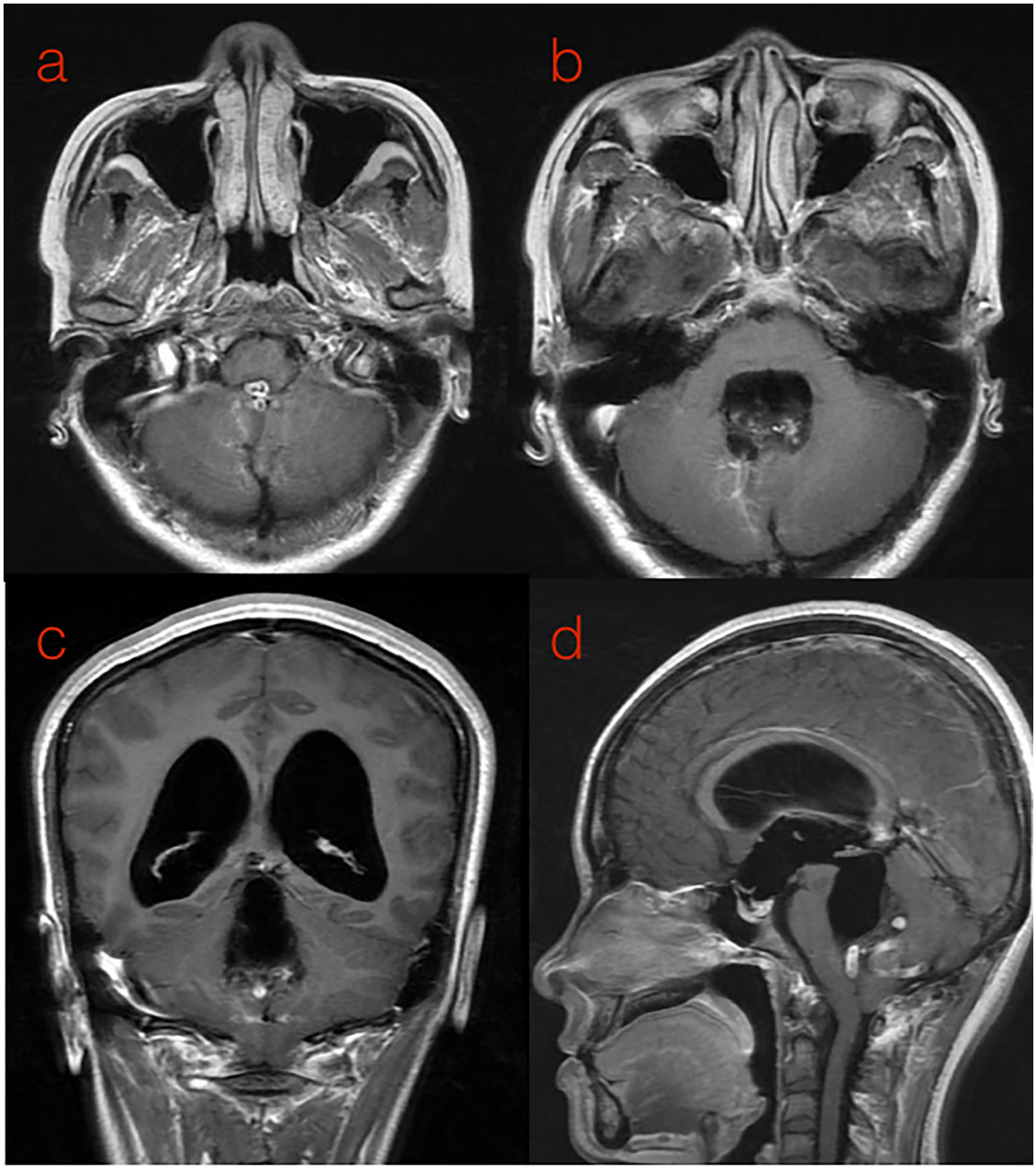

Fig. 1 – The upper column (a and b) showed contrast enhanced axial T1 weighed image, and the lower colum (c and d) showed contrast enhanced coronal and sagittal T1weighed image, separately. (a and d) Engorged and tortuous medullary veins fulfilled the outlet of fourth ventricle, at the level of occipital-cervical junction. (b–d) Diffusely dilated bilateral lateral ventricle, third ventricle, cerebral aqueduct Sylvius, and fourth ventricle, and right cerebellar developmental venous anomaly (DVA) with typical characteristics of ''caput medusa'' in (c).

3. Discussion

DVAs, accounting 63% of intracranial vascular malformation in autopsy series [1], are most common entity of the cerebrovascular malformation, and mostly found incidentally by brain computed tomography (CT) and magnetic resonance imaging (MRI). It is considered as a normal variation of the cerebral venous angioarchitecture rather than a pathological disease, with a cluster of transmedullary veins converging into a large caliber collecting vein, and further drain in the superficial or deep venous system after crossing the brain parenchyma. Most DVAs are asymptomatic, however, there are still complications caused by DVAs itself or accompanied vascular lesion, inclusive of parenchymal hemorrhage, venous

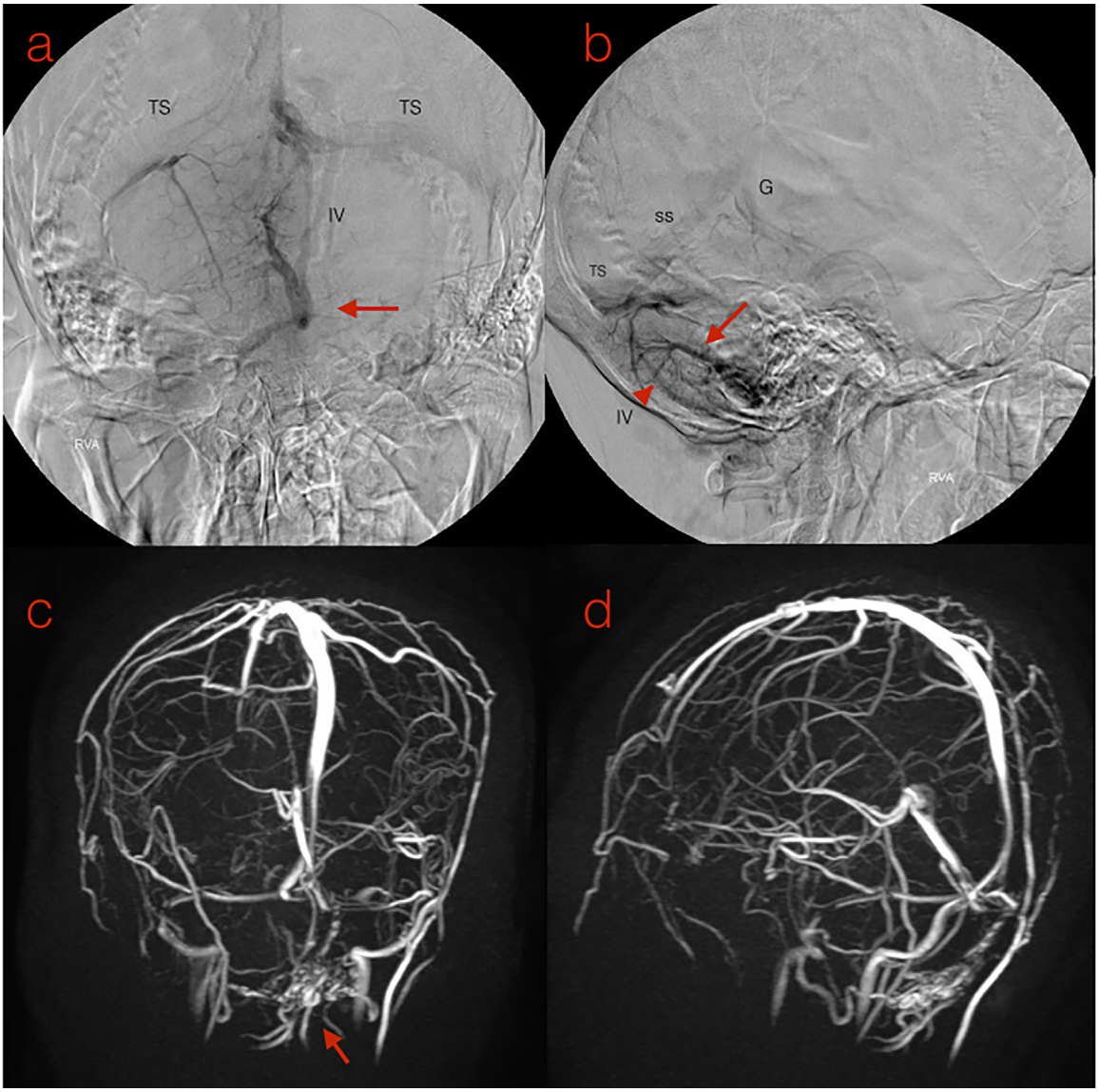

Fig. 2 – The upper column showed vertebral angiogram, venous phase. The lower column showed magnetic resonance venography (MRV). TS: transverse sinus; IV: inferior vermian vein; SS: straight sinus; G: vein of Galen. (a) AP and (b) lateral. DVA with engorged medullary vein over the floor of fourth ventricle, with collecting vein drain in the inferior vermian vein and further into the straight sinus, with typical caput medusae pattern under vertebral angiogram. (c and d) Three-dimensional reconstructive MRV showed engorged tortuous medullary veins over the floor of fourth ventricle, compatible to previous MRI findings.

ischemic infarction, subarachnoid hemorrhage, intraventricular hemorrhage, obstructive hydrocephalus caused by the aqueduct Sylvius compression, and nerve root compression [4,5]. Pereira et al. indicated that symptomatic DVAs can be further categorized into three types, mechanical (14/69 patients), flow-related (49/69 patients) and idiopathic (6/69 patients) types underlying DVAs, and obstructive hydrocephalus belongs to the mechanical type. From their series, obstructive hydrocephalus (50%, 7/14 patients) is the most common finding in mechanical type, and the most common compressed site is the aqueduct Sylvius (6/7 patients with obstructive hydrocephalus) [4].

For our patient, there is solely right cerebellar DVA without other related vascular lesion, such as cavernous malformation (CM), superficial venous malformation of the head and neck or other vascular malformation [5], and with presentation of obstructive hydrocephalus of the outlet of fourth ventricle.

After reviewing English literature of obstructive hydrocephalus with DVAs, there are twelve cases series nowadays and all related to the compression of the aqueduct Sylvius [2]. For our patient with fourth ventricle outlet obstruction caused by cerebellar DVA, it is a rare entity and not found in previous literature. From brain DSA and MRV of our patient, we found that engorged transmedullary veins of the right cerebellar DVA

obscured the floor of fourth ventricles, and then drained centripetally and radially into the collecting vein, and then drain in the inferior vermian vein and finally terminate at the straight sinus (Fig. 2).

Patients with DVAs mostly are benign and further followed up imaging series or treatment are not necessary. However, in patients with symptomatic DVAs, the further management should be made after careful deliberations. For those with symptomatic DVAs coexisting vascular malformation, precise surgical extirpation of the coexisting vascular malformation rather than DVAs itself is warranted. For symptomatic DVAs solely without other vascular lesion, complications management rather than DVAs itself is warranted, such as alleviating the mass effect from intracerebral hemorrhage, decompressive craniectomy for extensive cerebral infarction or edema, and cerebrospinal fluid (CSF) diversion for ventricular outflow obstruction. Extirpation of the DVAs is not recommended owing to catastrophic venous infarction or brain edema after interrupting normal venous drainage system [3]. For our patient with obstructive hydrocephalus due to fourth ventricle outlet obstruction, cerebrospinal fluid diversion is the option of treatment.

Herein, we reported a rare case of symptomatic DVA obstructing the outlet of fourth ventricle with subsequent hydrocephalus, and ventriculo-peritoneal shunt proved effective to improve the patient's outcome.

4. Conclusions

Developmental venous anomalies are common entities in cerebral malformation, and symptomatic DVAs need to be evaluated precisely. Obstructive hydrocephalus induced by DVAs is infrequent and not caused solely from the aqueduct Sylvius obstruction. Any lesion involved in the cerebrospinal fluid circulation pathway should be evaluated carefully, such as the fourth ventricle outlet obstruction in our patient.

Conflict of interest

All authors have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.

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