Clinical Case Report

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Obstructive hydrocephalus associated with spontaneous dural arteriovenous fistula

A case report

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Abstract

Rationale: Dural arteriovenous fistulae (DAVF) are vascular disorders depicted with direct interconnection between dural arteries and cerebral venous sinuses and/or cortical veins. Only a hand full of cases have been reported in literature. DAVF obstructing the 3rd ventricle and quadrigeminal cistern resulting in hydrocephalus is very rare.

Patient concerns: We present a 55-year-old female with 2 years history of headaches and blurring of vision. Cranial nerves examinations were unremarkable.

Diagnoses: Magnetic Resonance Imaging (MRI) and digital subtraction angiography showed multiple tortuous vascular malformation in the 3rd ventricle and quadrigeminal cistern resulting in obstructive hydrocephalus (OHC).

Interventions: We utilized endovascular embolization treatment option to obliterate the lesion. We used Onyx embolization agent (ev3 Neurovascular, Irvine, CA) to embolize the lesion via the transarterial route.

Outcomes: The OHC resolved spontaneously after the endovascular embolization of the DAVF. The patient recovered with no further neurologic complication. Two years follow-up reveal no recurrence of the DAVF as well as hydrocephalus.

Lessons: Adequate knowledge on the vascular anatomy is very crucial in managing DAVF.

Abbreviations: CTA = computed tomography angiography, DAVF = dural arteriovenous fistulae, DSA = digital subtraction angiography, MMA = middle meningeal artery, MRA = magnetic resonance angiography, MRI = magnetic resonance imaging, OHC = obstructive hydrocephalus.

Keywords: arteriovenous, dural, embolization, hydrocephalus, malformations

1. Introduction

Dural arteriovenous fistulae (DAVF) are vascular disorders depicted with direct interconnection between dural arteries and cerebral venous sinuses and/or cortical veins.[1,2] Most DAVF occur at the transverse, sigmoid, as well as the cavernous sinuses. Nevertheless, cases of DAVF have been reported at different locations within the dura mater.[1,2] Majority of DAVF reported in literature are idiopathic. However, a hand full are linked to antecedent procedures like craniotomy, traumatic events, as well as thrombosis of the dural sinus.[3,4] Furthermore, cases of parenthetical DAVF have also been reported. In such cases, the signs and symptoms were directly associated with site and pattern of venous drainage.[3,5] We present a rare case of DAVF associated with obstructive hydrocephalus (OHC).

2. Case report

A 55-year-old female presented with a 2-year history of headache. She also experienced progressive blurring of vision. Cranial nerves examinations were unremarkable. General systemic examination was also unremarkable. Preoperative computed tomography (CT) scan revealed hematoma at the midbrain with enlarged right and left lateral ventricles (Fig. 1A–C). Magnetic resonance imaging (MRI) showed multiple tortuous vascular malformation in the 3rd ventricle and quadrigeminal cistern resulting in OHC (Fig. 2A–D). Digital subtraction angiography (DSA) confirmed that the arteriovenous malformation was DAVF (Fig. 3A). The DAVF was fed by external carotid artery, posterior cerebral artery as well as the middle meningeal artery (MMA) and flowed into the vein of Galen. There were no other associated lesions like tumors during our evaluation of the patient. Routine laboratory investigations were unremarkable. Chest X-ray and ECG were also unremarkable.

We utilized endovascular embolization treatment option to obliterate the lesion. Embolization was done via the transarterial

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The ethical committee of West China Hospital full approved our case study. The patient and her relatives were informed about our intension to involve her in a case study and they agreed to partake in the study. They signed the concern form before the operation was carried out according to all surgical protocols.

The patient and her relatives were dually informed about our intention to publish the case and they fully concerted to the use of his documents. A written informed consent was obtained from the patient. The hospital also concerted to the use of his information for publication.

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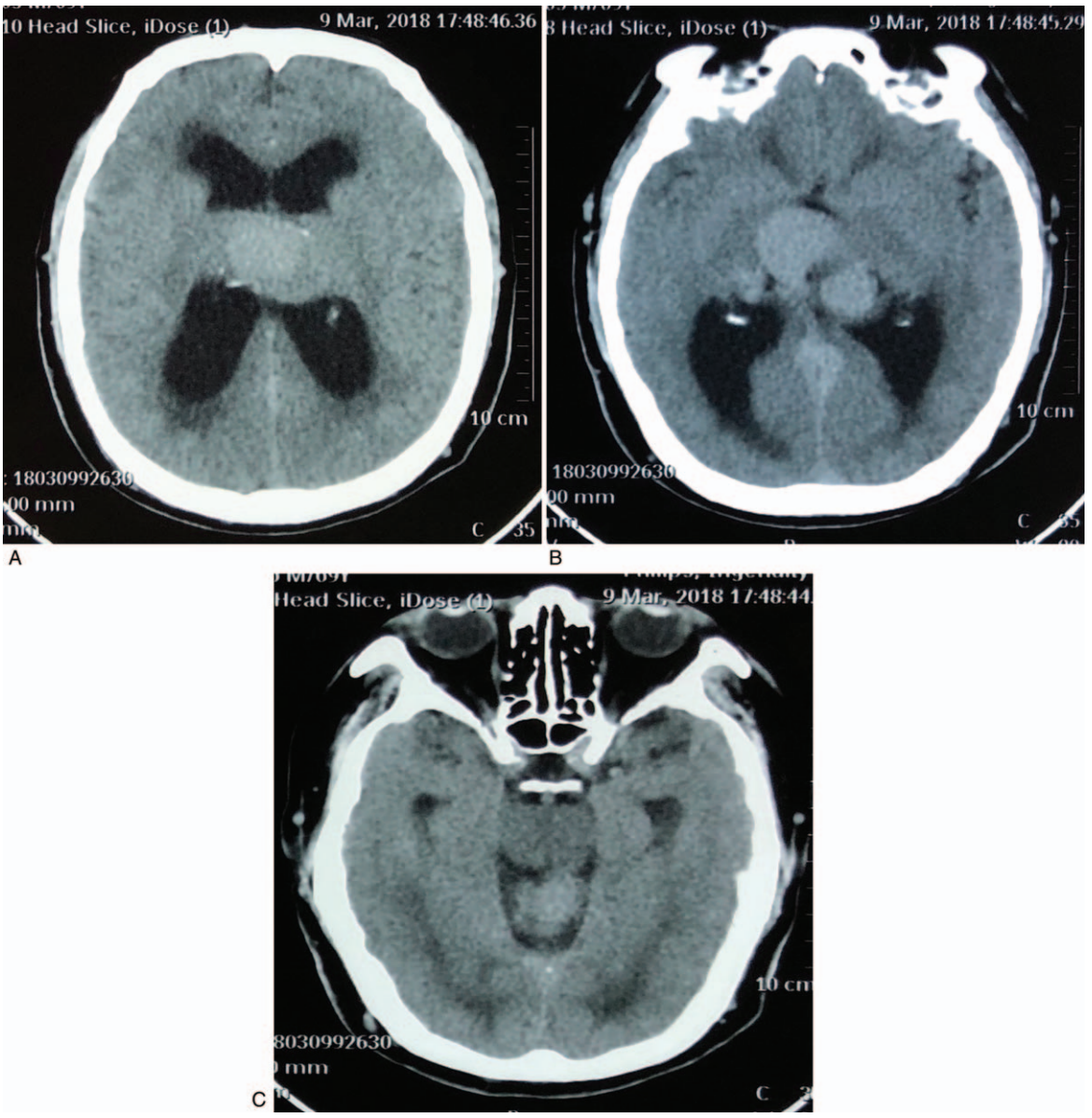

Figure 1. (A–C) Preoperative computed tomography scan images showing hematoma at the midbrain.

route. We 1st introduced the microcatheter into the MMA and then embolized the lesion with Onyx (ev3 Neurovascular, Irvine, CA). Subsequently, we introduced the microcatheter into the posterior cerebral artery and then embolized this branch too with Onyx (ev3 Neurovascular). We notice partial resolution of the OHC after endovascular embolization of the DAVF a few days after the operation and complete resolution on follow-ups. Postoperative radiologic examination showed DAVF disappeared (Fig. 3B, C). Two years follow-up revealed no recurrence of the DAVF as well as hydrocephalus. She is currently well and goes about her normal daily activities.

3. Discussion

The DAVF are curable vascular disorders that are associated with direct interconnection between dural arteries and cerebral venous sinuses and/or cortical veins. These lesion forms about 10% to 15% of all vascular malformations in the brain.[6,7] Most patients with these lesions are in their 50s and 60s. So far, no sex dominates have been associated with DAVF.[3,6] Also, no genetic or family links has been associated with these lesions. It has been demonstrated that majority of DAVF are idiopathic.[3] Our case was a spontaneous occurrence. Nevertheless, some occurrences have been linked to antecedent procedures like craniotomy, traumatic events, and thrombosis of the dural sinus.[3]

Headaches, vertigo, tinnitus, as well as neurologic deficits are most prominent clinical features of DAVF.[5,6,8,9] In our case, however, apart from the above symptomatology, she also experienced progressive blurring of vision. In more severe cases, there is usually associated fatal intracranial hemorrhage.[6,8] Also, increased venous pressure usually leads to increased intracranial pressure (ICP) in some patients with DAVF.[1] Moreover, hydrocephalus is rarely seen in patients with DAVF.[1,9] However, hydrocephalus is seen in about 28% of cases with tentorial DAVF.[10] Hydrocephalus in patients with

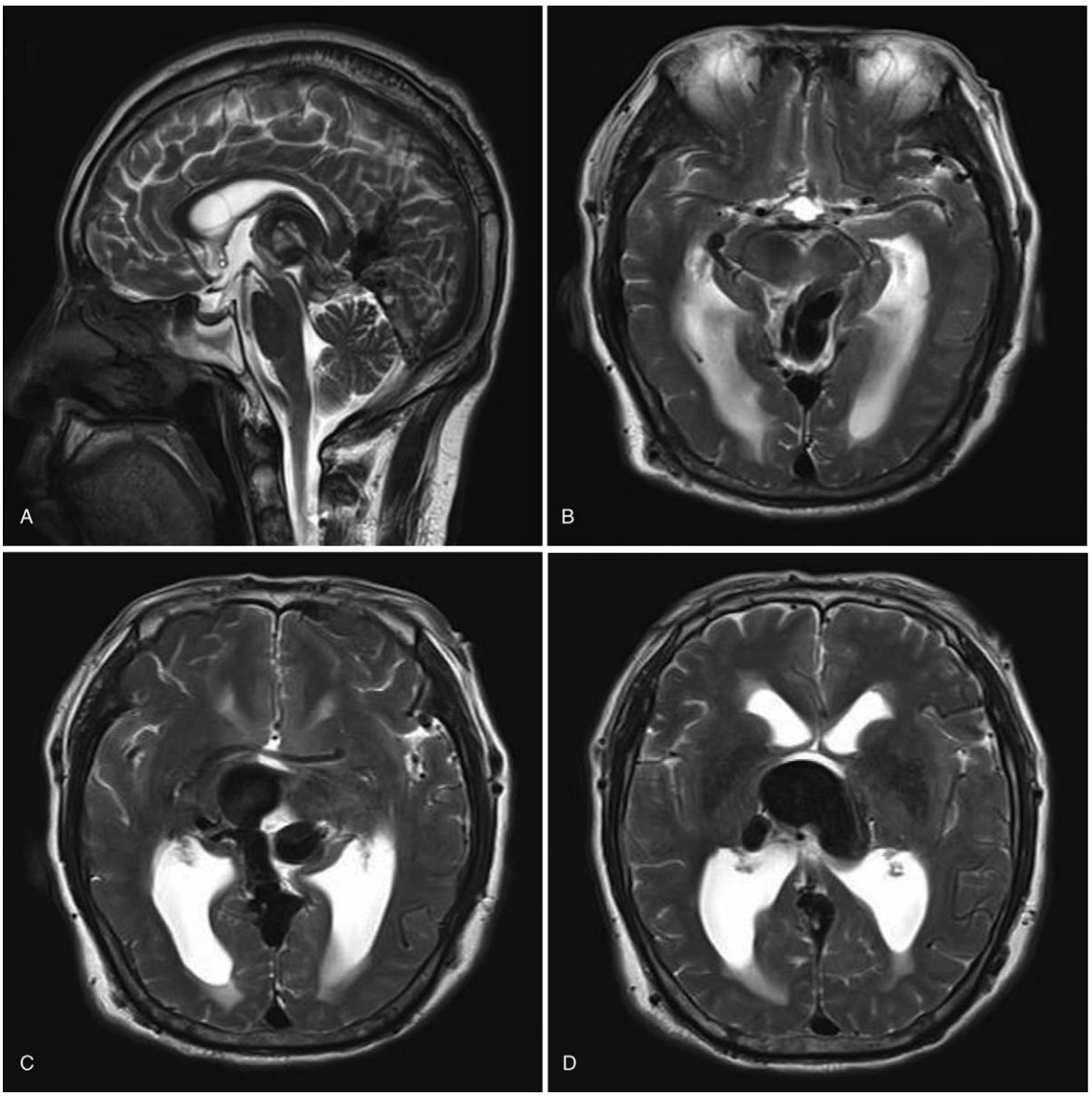

Figure 2. (A–D) Preoperative magnetic resonance imaging showing multiple vascular malformations located in the 3rd ventricle and quadrigeminal cistern. (A–C) flow-empty effect observed in T2-weight series. (D) Dilated veins blocking the 3rd ventricular outflow tract leading to obstructive hydrocephalus.

DAVF is most likely due to compression of the cerebral aqueduct by venous varices.[1]

Ernst and Carlson reported a case of DAVF with OHC.[1] They indicated that the cause of OHC in their case was as result of colossal congestion of the posterior fossa leading to massive reflux into the intraparenchymal venous system.[1] The OHC in our case was as a result of obstruction of the 3rd ventricle and quadrigeminal cistern by multiple tortuous vascular malformation (Fig. 2A–D). In addition, van Buggenhout et al presented a case of DAVF with OHC in whom the mechanism of hydrocephalus was as a result of engorgement of the vein of Galen.[11] Nevertheless, in their case, the DAVF was located in the quadrigeminal cistern just similar to our case. The whole malformation cause obstruction of the cerebral aqueduct.[11] Furthermore, some authors observed similar cases of DAVF with OHC in whom the hydrocephalus was as a result of drainage into the deep venous system leading to dilation of the vein of Galen.[1,10] In addition, Tsugane et al reported a similar case with engorgement of the straight sinus resulting in blockade at the junction of the cerebral aqueduct and 4th ventricle.[12] Satoh et al reported 4 cases of DVAF with cerebellar hemorrhage adjacent the 4th ventricle with associated OHC.[13] Kataoka and Taneda also reported a case of OHC as result of minor hemorrhage in the lateral and 3rd ventricles that resolved spontaneously.[14] Our case was also associated with hematoma at the midbrain.

The gold standard modality of diagnosing and classification of DAVF is catheter angiography.[3,15–17] Nevertheless, modern diagnostic modalities like CT angiography (CTA) and MR angiography (MRA) have further improved the detection of these malformations.[3,18,19] Furthermore, CTA is much more usually in surgical planning. CTA is able to differentiate the draining veins from other brain structures.[3] On the contrary, MRA is able to identify flow voids from draining veins as well as T2 hyperintensity from preceding ischemia triggered by venous hypertension.[3] Our initial radiologic assessment method was CT scan and MRI. MRI showed multiple tortuous vascular

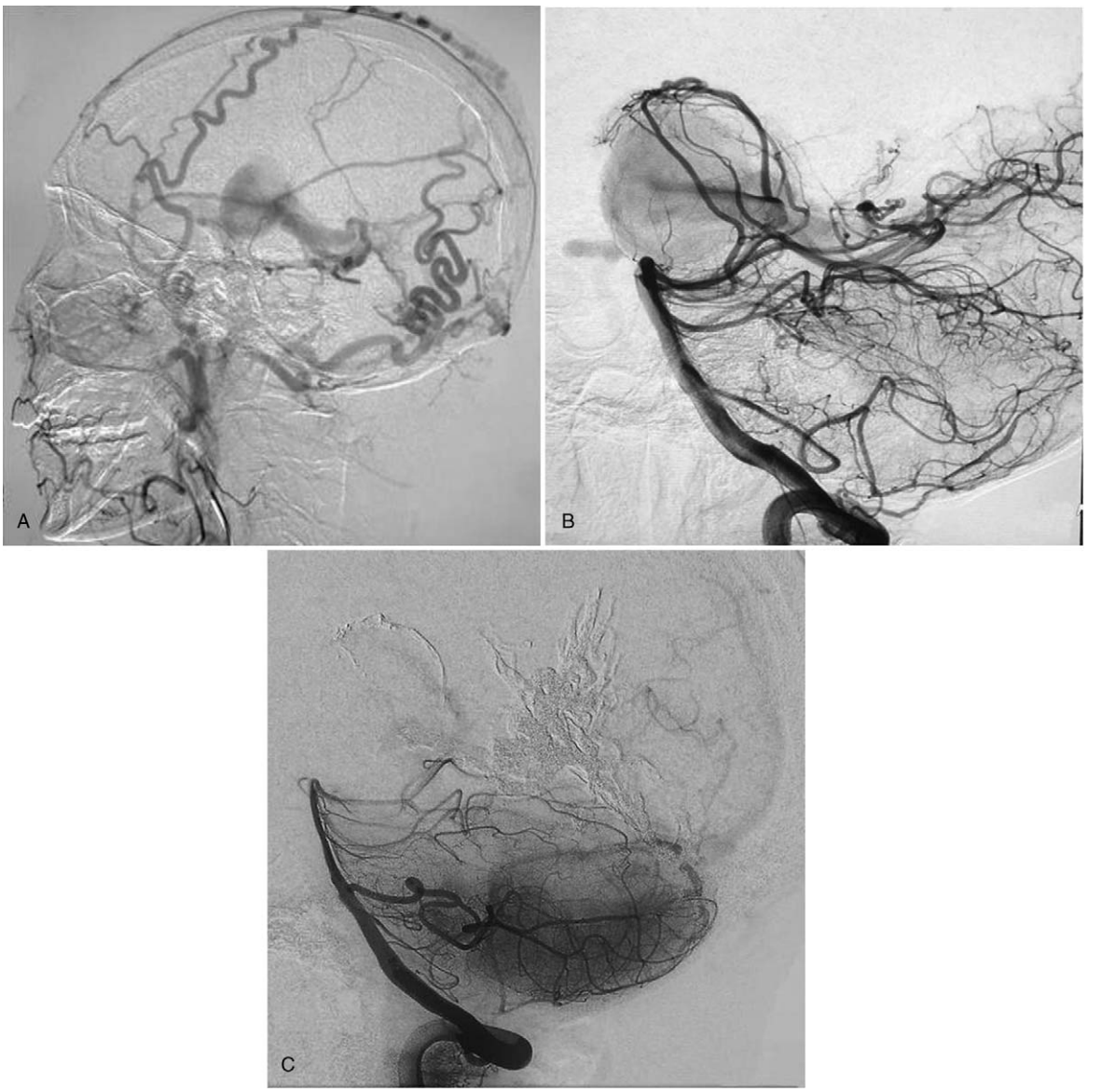

Figure 3. (A) A preoperative digital subtraction angiography (DSA) image showing the vascular malformation (DAVF). (B, C) Postoperative DSA images showing disappearance of the DAVF after endovascular embolization treatment.

malformation in the 3rd ventricle and quadrigeminal cistern resulting in OHC. CT scan also revealed hematoma at the midbrain.

We utilized DSA to confirm that the lesions were arteriovenous malformation and specifically DAVF.

Dural AVFs have been successfully treated by endovascular occlusion and/or surgical intervention.[10,13] Traditionally, essential surgical skeletonization of the affected sinus is very rewarding in most cases of DAVF.[13] Endovascular procedures are usually via transarterial or transvenous embolization or both. In most cases with high-risk fistulae, endovascular treatment with Onyx is satisfactory.[1] In our case, we utilized endovascular embolization treatment option to obliterate the lesion. Embolization was done via the transarterial route. In cases with hematoma at the midbrain, the theory of increasing hematoma volume with time is utilized during emergency evacuation of the hematoma followed by excision of the malformation.[13] The associated hematoma at midbrain resolved during the management of our case. In some patients, clipping of the retrograde venous drainage is an efficient technique in managing DAVF.[13,20] We did not have any limitations during the management of this case.

4. Conclusion

The OHC associated with DAVF is extremely rare. In our case, the DVAF was seated deep in the brain. OHC was as a result of obstruction of the 3rd ventricle and quadrigeminal cistern by multiple tortuous vascular malformation. We successfully obliterate the DAVF using endovascular embolization. The OHC resolved spontaneously after the endovascular embolization. The patient recovered with no further neurologic complication. Two years follow-up reveal no recurrence of the DAVF as well as hydrocephalus.

Author contributions

All the authors conceived the project and designed the study. CWZ, WF, SAR and HS collected patient's data. XDX provided technical assistance in the study. SAR prepared the illustrations and wrote the paper. All authors approved the paper for the submission.

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