Multiple intracranial dural arteriovenous fistula

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Dural arteriovenous fistula (DAVF) is also known as dural arteriovenous malformation. Two forms of DAVF have been introduced, however, here we present an exceptional case of DAVF with unique origin and drainage. In this study, we present a rare case of multiple DAVFs in a 50 year old man with right parietal intraparenchymal hemorrhage. MRI showed two round right parieto-occipital masses with flow void intensity adjacent to superior sagittal sinus (SSS). Another pathology connected to SSS by an abnormal cortical vein was detected anterior to first lesion. This study showed that both DAVFs were simultaneously drained in SSS in our patient.

Key words: Arteriovenous malformation, multiple dural arteriovenous fistula, multiple lesions

INTRODUCTION

Intracranial dural arteriovenous fistulas (DAVFs) are abnormal arteriovenous shunts developing on the dura mater, usually within the wall of a dural sinus, which can be asymptomatic for years.[1-9] Majority of DAVFs are idiopathic and solitary.[1,5] Intracranial DAVFs are fairly common, accounting for 10-15% of all intracranial arteriovenous shunts,[6] whereas multiple DAVFs at separate sites are relatively rare, accounting for 7% of all intracranial DAVFs.[10-13] Patients with multiple DAVFs usually present with life-threatening hemorrhages related to venous outflow obstruction or acute dysfunction. The exact mechanism of occurrence and natural history remain undetermined, but various factors, such as congenital development, venous hypertension, head trauma, venous thrombosis, and radiation therapy, may be involved.[7,10,14]

Multiple DAVFs, although rare, do exist and it is important to look for any evidence of their presence when evaluating patients with symptoms suggestive of arteriovenous fistulas. In this study, we present a case of multiple DAVFs. We have focused on this unique type of venous drainage in dural arteriovenous fistulas.

CASE REPORT

A fifty year old man with history of long term headache and sudden onset of decreased level of consciousness was admitted to Namazee Hospital, affiliated to Shiraz University of Medical Sciences, Shiraz, Iran, in 2011. Brain computed tomography (CT) showed right parietal intraparenchymal hemorrhage [Figure 1]. The patient had glasgow coma scale (GCS) 12 on arrival.

Brain magnetic resonance imaging (MRI) with and without gadolinium was performed to rule out tumor apoplexy that showed two round right parieto-occipital masses with flow void intensity adjacent to superior sagittal sinus (SSS) [Figure 2]. Brain CT angiography (CTA) revealed a mass of $3 \times 2 \times 2$ cm connected to SSS in posterior part of right parieto-occipital area. Another pathology connected to SSS by an abnormal cortical vein was detected anterior to first lesion [Figure 3]. Digital subtraction angiography of brain vessels showed large vascular pathology that filled at early arterial phase by a torturous extra cranial vessel and drained into the posterior third of SSS. More anterior lesion was smaller vascular pathology that supplied at early arterial phase by posterior branch of middle meningeal artery and drained into SSS by an abnormal cortical vein that classified according to Borden classification type II [Figure 4].[15]

Regarding the patient's history and images, DAVF was diagnosed. Because of the site of his pathologies, multiplicity and difficulty of endovascular treatment, open craniotomy was suggested for removal. General anesthesia was administered in prone position and the face rotated to the left side. In this position right parieto-occipital area was in highest part. Then a large right parieto-occipital craniotomy with excellent exposure of SSS was performed. When the bone was elevated, bleeding occurred. Source of bleeding was from two dilated meningeal arteries that was controlled easily. Dura was opened and the two large masses with separate arterial and venous structure were found. The larger pathology had a large vascular pedicle that had connection to SSS and dural vessels and was ligated [Figure 5]. After preparation of the site for applying emergency temporary clip, the thick wall mass
that was free of pulse was dissected carefully. When the lesion was removed from parenchyma, a permanent titanium yasargil clip was applied on its vascular pedicle and the lesion was resected totally without parenchymal damage. The anterior pathology was removed in the same manner. Abnormal cortical vein was also resected. Hemostasis was carried out and dura was closed.

Postoperative brain CT-scan of the patient was acceptable and there was no vascular pathology in the brain CTA. No neurological deficit was found. The patient was discharged from the hospital with GCS 14.

DISCUSSION

Intracranial DAVFs with retrograde cortical venous drainage are aggressive lesions presented by intracranial hemorrhage, intracranial hypertension, seizures, progressive neurological deficits, and dementia. Prompt treatment is necessary because the natural history of untreated lesions is associated with a poor prognosis.\textsuperscript{[1,2,3]}
Intracranial DAVFs can be classified according to the type of venous drainage. Borden, et al.[15] proposed a classification of DAVFs, grouped into three types based upon their venous drainage including type I with drainage to a dural sinus, type II with drainage to a dural sinus and reflux into cortical veins, and type III only with drainage to the cortical veins. Aggressive clinical presentation correlates strongly with Borden types: 2% of type I, 40% of type II, and 80% of type III.[5,13]

DAVFs are presented with intracranial hemorrhage or neurological deficits. However, source of bleeding was controlled easily in our study. Treatment of DAVFs with cortical venous drainage is aimed in occlusion of the venous drainage or all arterial supply that can be surgical, endovascular, or a combination of both.[4‑16]

Most DAVFs are idiopathic and solitary.[13] The term multiple DAVF has two different meanings in literature. Some authors use the term to indicate DAVFs that developed simultaneously at different sinuses, whereas the others use it to indicate a second DAVF that developed at other sinuses at the interval after resolution of initial DAVF.[5] The presence of two or more different DAVFs draining into one single sinus has not been described. In this case, both DAVFs simultaneously drained into the SSS in our patient. Both DAVFs were connected to two abnormal giant vascular pockets that developed intraparenchymal hemorrhage and were different to a simple DAVF or the multiple DAVFs described elsewhere.

Based on the findings within this case, we suggest that the definition and classification of DAVFs be revised, and a new category including separate lesions draining into a common venous drainage be considered. Such a classification scheme of course needs report and collection of further cases into a comprehensive case series.

CONCLUSION

Planning of treatment strategies for multiple DAVFs requires careful analysis of the venous drainage from the affected sinuses and cerebral hemodynamics. We presented a unique type of multiple DAVFs draining into a common venous drainage SSS instead of the routinely reported separate drainage. The introduction of this new case opens a new concept, which may be useful for further classification or suggestion of the responsible pathophysiologic processes.

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