THE ASEAN JOURNAL OF RADIOLOGY 2010; 16(1): 77-84 77

Original Article

Malignant Cranial Dural Arteriovenous Fistula with Symptom Aggravated by Spontaneous Venous Thrombosis

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Introduction

Intracranial dural arteriovenous fistulae (DAVF) is a disease with a variety of presenting symptoms, depending on the shunt location and venous drainage route, and multiple treatment strategies such as transcatheter embolization, surgical clipping, radiation therapy.[1,2] The disease is considered an acquired disease initiated by thrombosis of the intracranial venous system mostly the dural sinuses and later on developed arteriovenous fistulae at the level of the dural mater. Spontaneous regression of the DAVF has been widely reported, which is likely caused by thrombosis of the sinus or fistula.[3] Spontaneous thrombosis of the venous outlet route can occur before closure of the fistula and causes worsening of the patient's symptom, such instance frequently occurred with the cavernous DAVF with spontaneous thrombosis of the superior ophthalmic veins (SOV). Up to date, spontaneous thrombosis of the cerebral veins in cases of cranial malignant DAVF has rarely been reported as well as the role of anticoagulant as an additional treatment.

Purpose

To report two cases of malignant intracranial DAVF whose symptoms worsen by spontaneous thrombosis of the CVR.

Method

We reported two cases of malignant cranial DAVF with spontaneous thrombosis of the cerebral vein presented at our institute during April 2009 to June 2009 (1 male 22-year-old; 1 female 62-year-old), in terms of presenting symptoms, imaging features, therapeutic methods, and clinical outcomes.

Illustrative cases

Patient 1: A 22-year-old man presented with headache and repeated seizure for 2 months. He suddenly developed deteriorated conscious level and was sent to our hospital. The initial blood workup which included CBC, electrolytes, renal function and coagulogram were normal. The brain CT scan showed multiple dilated and tortuous hyperdense supratentorial veins which suggestive evidence of

DAVF with CVR. The patient's symptoms were suspected due to intolerance of the brain to chronic venous congestion from the CVR. Cerebral angiogram showed DAVF at the middle 1/3 of the superior sagittal sinus (SSS) with arterial supply from bilateral middle meningeal arteries, transosseous branches of both superficial temporal arteries, and pial supply from the cortical branches of both ACAs. There were CVR into both cerebral hemispheres superficial cortical veins and occlusion of the anterior 1/3 of the SSS and right transverse sinus. Transarterial embolization with n-butyl-2-cyanoacrylate (NBCA) was done with significant reduction of the shunt flow and CVR. Three days post embolization, there was no significant improvement of the patient's conscious level. The patient's history, brain CT scan, angiograms, and the follow up brain MRI (figure 1 and 2) were then reviewed and found that massive thrombosis of the deep venous system was missed diagnosed initially and worsening of the patient's neurologic symptoms were likely to be aggravated by thrombosis of the deep cerebral veins. Low molecular weight heparin was given with improvement of the patient's neurological status.

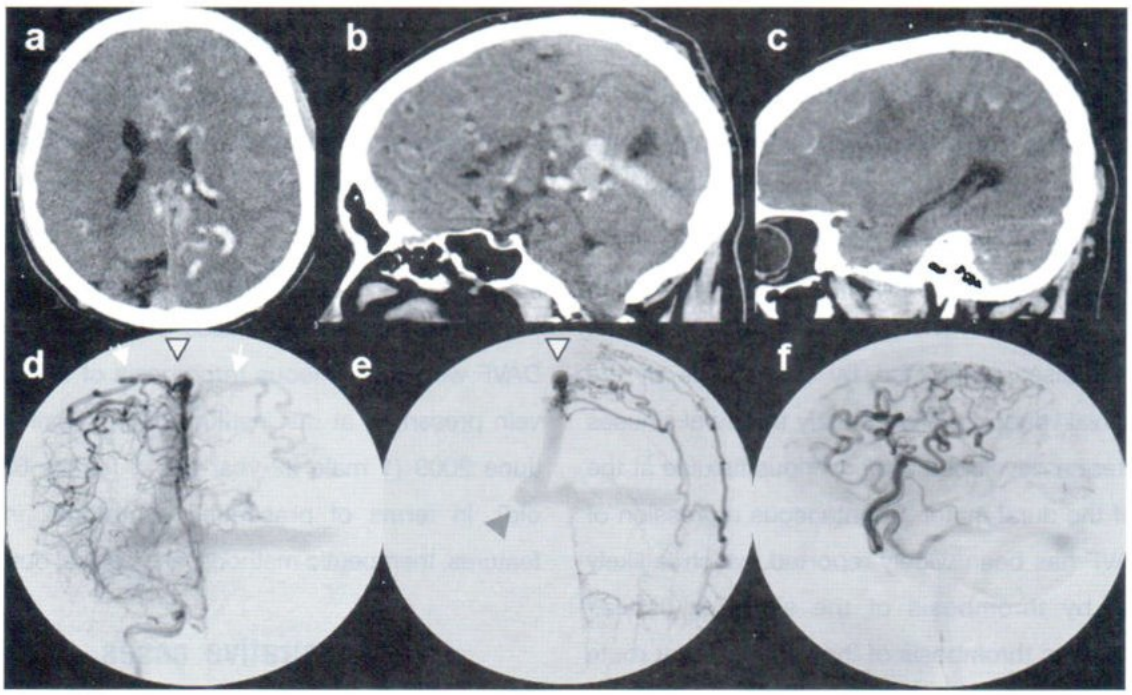

Fig.1 Non contrast brain CT scan axial (a) mid-sagittal (b), right parasagittal views (c) showed multiple hyperdense vessels at the midline and left parasagittal region with hyperdensities within the Galenic vein and straight sinus. Tonsilar brain herniation was evident in the mid sagittal veiw. Patchy hypodensities were seen at the right cerebrum white matter as well as curvilinear subcortical calcifications, a finding of chronic venous congestion. Right ICA angiogram late arterial phase (c) and left external carotid angiogram (d) AP view showed DAVF at the middle 1/3 of the SSS (arrow heads) with occlusion of the right transverse sinus (read arrow) and bilateral CVR (yellow arrows). Left ICA angiogram lateral view capillary phase (f) showed the appearance and drainage route of the left-sided CVR.

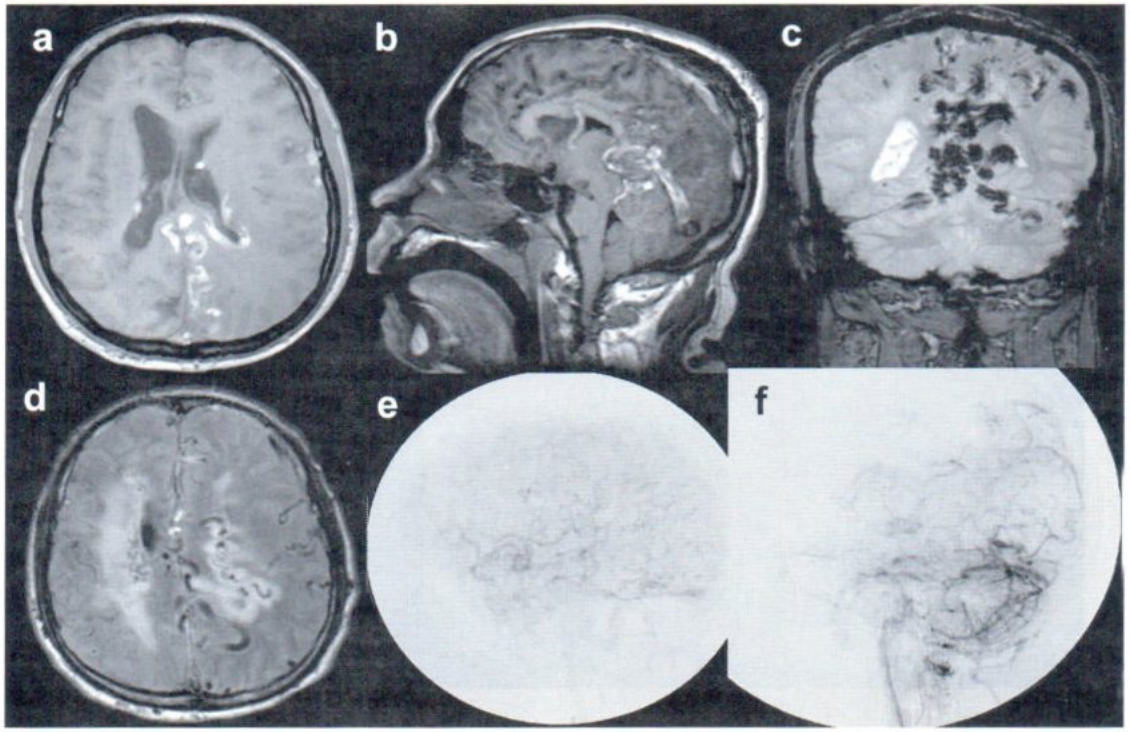

Fig.2 Post embolization MRI noncontrast axial TIWI (a), sagittal T1WI (b), and coronal GRE T2* showed massive thrombosis of the cerebral veins and deep venous system evident by T1 hyperintense signal within the vessel lumens and hypointense signal blooming on GRE T2*. Axial FLAIR (d) showed extensive T2 hyperintense signal at both cerebral hemispheres white matter. Retrospective reviewed of the late venous phase right ICA (e) and vertebral (f) lateral view angiograms showed cerebral venous congestion and no visualization of the deep venous system, confirming evidence of venous thrombosis preembolization.

Patient 2: A 62-year-old female presented with acute onset of nausea and vomiting and deteriorated conscious level later on. The brain CT scan showed an acute intracerebral hematoma (ICH) at the perinsular region of the right frontal lobe, enlarged bilateral cavernous sinuses and superior ophthalmic veins (SOV), and dilatation of the cortical veins at the right frontal and temporal lobes. Cavernous DAVF with CVR was suspected and the bleeding was thought due to hemorrhagic venous infarction from the CVR. Urgent cerebral angiogram showed DAVF at both cavernous sinuses with venous reflux into both superior ophthalmic veins (SOV) and CVR into the right superficial middle cerebral vein. Transvenous coil embolization was done with complete obliteration of the CVR and nearly complete obliteration of bilateral cavernous DAVF flow. Three days post embolization, the patient neurologic symptom worsen by developed grade IV left-sided weakness. Follow up brain CT and MRI showed worsening of the hemorrhagic venous infarction and thrombosis of two right intrasylvian veins. Retrospective reviewed of the initial brain CT scan showed that thromboses of the right intrasylvian veins were already evident and miss diagnosed. Low molecular weight heparin (LMWH) was given with

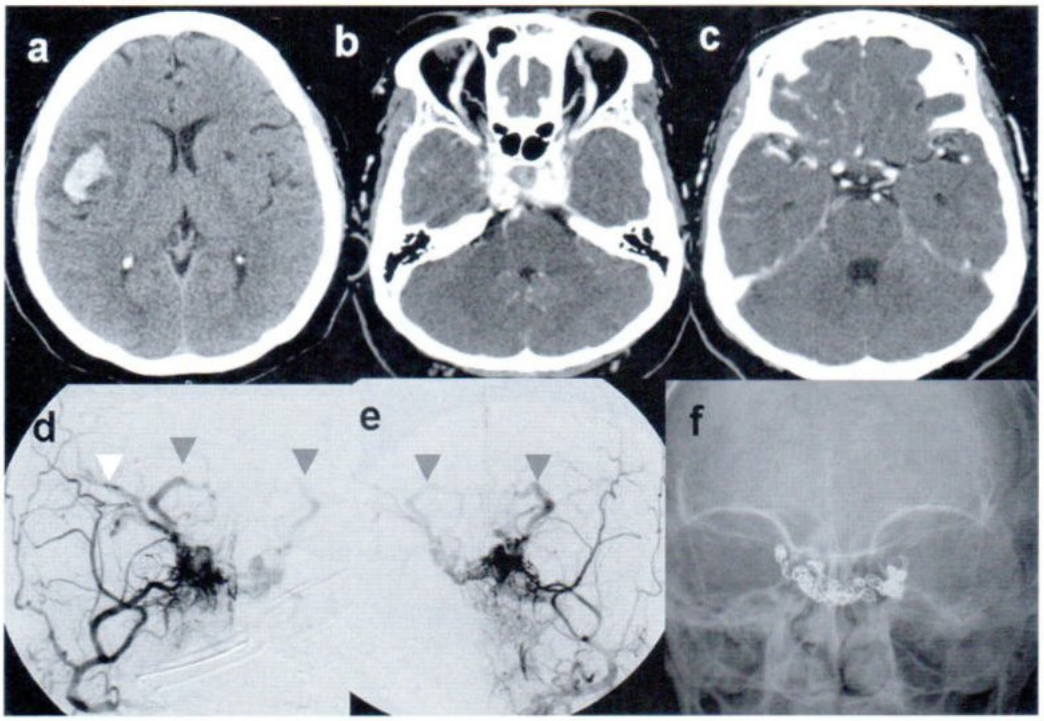

Fig.3 Noncontrast brain CT (a) showed an acute ICH at the right frontal lobe periinsular region and an old lacune at the left lentiforn nucleus. Contrast enhanced CT showed (b,c) showed enlargement of both cavernous sinuses, SOVs, and too much enhanced vessels at the right frontal and temporal lobes) (red arrows). Right external carotid (d) and left external carotid (e) angiograms AP view showed DAVF at both cavernous sinuses with venous reflux into the SOVs (green arrows), right middle cerebral vein (yellow arrow), and also the intercavernus sinus. Skull film AP veiw (f) showed the position of the transvenous embolized coils.

improvement of patient's neurodeficit and venous infarction on follow MRI.

Discussion

The association of venous thrombosis and cranial DAVF has been well known, mostly by the postulation that venous thrombosis occurs initially and then develop DAVF later on which could be seen by sequential follow up imaging. Recent studies have shown the evidence of thrombophilic abnormalities in patients with DAVF such as a higher level of D-dimer (a biological marker of endogenous fibrinolysis) and mutation of the prothrombin gene (G20210A).[4,5]

Cranial DAVFs with cortical venous reflux (CVR) are considered aggressive lesions. Aggressive symptoms include neurologic dysfunction related to intracranial hemorrhage or venous hypertension such as progressive dementia, seizures, and cerebellar symptoms. Thrombosis or partial thrombosis of the dural sinus with DAVF can often be demonstrated either by noninvasive vascular imaging or conventional angiography. However, symptomatic thrombosis of the cerebral vein in case of malignant cranial DAVF has rarely been reported. Demonstrable of intraluminal thrombus within the cerebral veins and continuous worsening of our two patient's neurologic symptoms despite disconnection of the

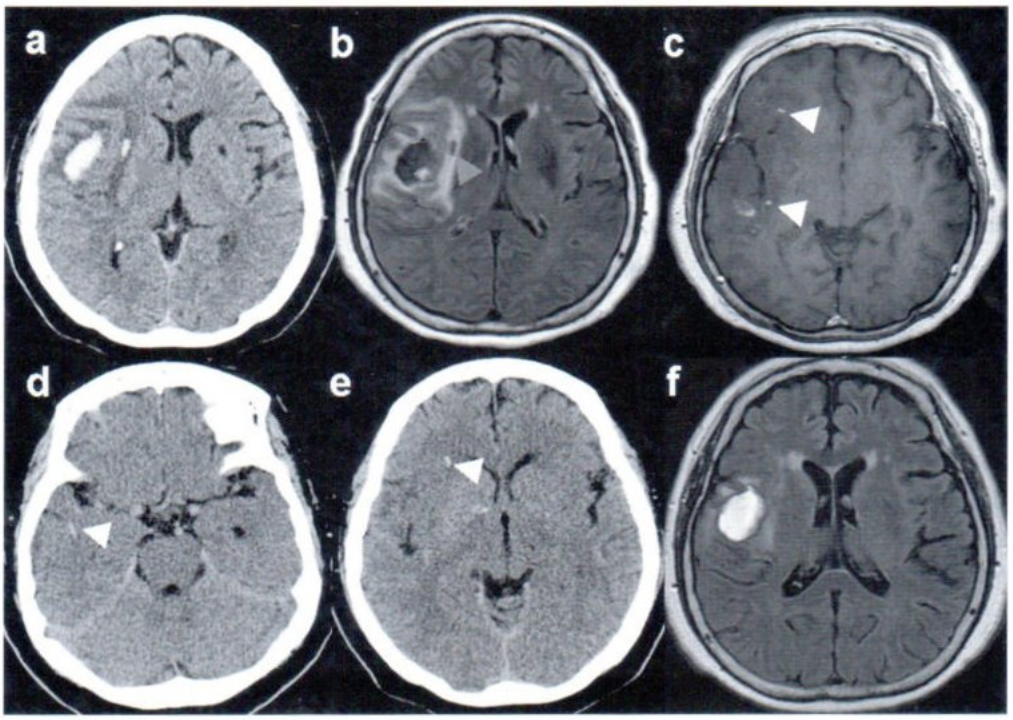

Fig.4 Noncontrast brain CT (a), MRI FLAIR sequence (b) showed expansion of the perihematoma brain swelling and a new small acute hemorrhage at the right external capsule (red arrows). MRI T1WI (c) showed two hyperintense thrombosed intrasylvian veins which was neglect on the initial brain CT (yellow arrows) (d, e). One month follow up MRI FLAIR sequence (f) after treatment with LMWH showed marked regression of the T2 hyperintense brain swelling and stable in size of the previous hematoma.

CVR by transcatheter embolization are the two main reasons for our postulation that thrombosis of the cerebral vein is actually the responsible cause for worsening of the patient's neurologic symptom and not by brain congestion from the CVR.

Anticoagulant was given as treating cerebral venoocclusive disease in both cases with continuous improvement of their symptoms on follow up. The role of anticoagulant in cranial DAVF has not been established. By our best searching of the medical data base, there are only a few reports of the usefulness of anticoagulant in cranial DAVF.[6,7] Therefore, thoroughly evaluation of the CT or MR images for evidence venous thrombosis in case of malignant cranial DAVF should always be performed because embolization alone may not improve the patient's symptom.

Conclusion

Associated cerebral venous thrombosis should be carefully searched in patient with malignant DAVF in pre-embolization stage, as even successful embolization alone may not improve clinical symptoms, and anticoagulant may play a major role as the parallel treatment to embolization.

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