Hong Kong J Radiol. 2011;14:39-43

CASE REPORT

A Rare Cause of a Large Posterior Fossa Mass: Giant Varix

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ABSTRACT

Herein, we report a very rare case of a patient suffering from the simultaneous occurrence of an unusually gigantic posterior fossa varix (measuring 35 mm x 45 mm) and a tentorial dural arteriovenous fistula in the absence of intracranial haemorrhage. Simultaneous occurrence of a varix and a tentorial dural arteriovenous fistula is present in only 4% of all intracranial dural arteriovenous fistulae. These concurrent conditions are associated with a high risk of intracranial haemorrhage and intracranial hypertension that can give rise to focal neurological deficits. Its clinical relevance and radiological findings are discussed in this report.

Key Words: *Cerebral hemorrhage; Cranial fossa, posterior; Hydrocephalus; Intracranial arteriovenous malformations; Varicose veins*

中文摘要

巨型靜脈曲張：後顱窩大腫塊的一個罕見病因

吳菀彬、劉國斌

本文報告一宗非常罕見的病例，患者有一個異常大的後顱窩巨型靜脈曲張（35 x 45毫米大小），同時有小腦幕硬腦膜動靜脈瘺（患者並無顱內出血）。硬腦膜動靜脈瘺患者中有4%會同時出現靜脈曲張及小腦幕硬腦膜動靜脈瘺。此異常情況與高風險顱內出血和顱內高壓有關，會導致神經系統局部功能受損。這宗病例是有關一個有異常大後顱窩靜脈曲張的病人，此患者並無顱內出血，但同時患有小腦幕腦腦膜動靜脈瘺。本文報告其放射性結果及有關的臨床意義。

INTRODUCTION

Dural arteriovenous fistula (DAVF) is a life-threatening neurological condition, which accounts for 10 to 15% of all intracranial arteriovenous lesions.[1] Simultaneous occurrence of varix and tentorial DAVF only occurs in about 4% of all intracranial DAVFs,[2] and is very rarely encountered in clinical practice. Although rare, the radiological features of both conditions are characteristic on magnetic resonance imaging (MRI) and angiography.

CASE REPORT

A 64-year-old man, with a history of smoking and hypertension, initially presented with left trigeminal and facial nerve disturbances. He was found to have an isolated left tentorial DAVF but refused treatment at

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Submitted: 2 Feb 2011; Accepted: 1 Mar 2011.

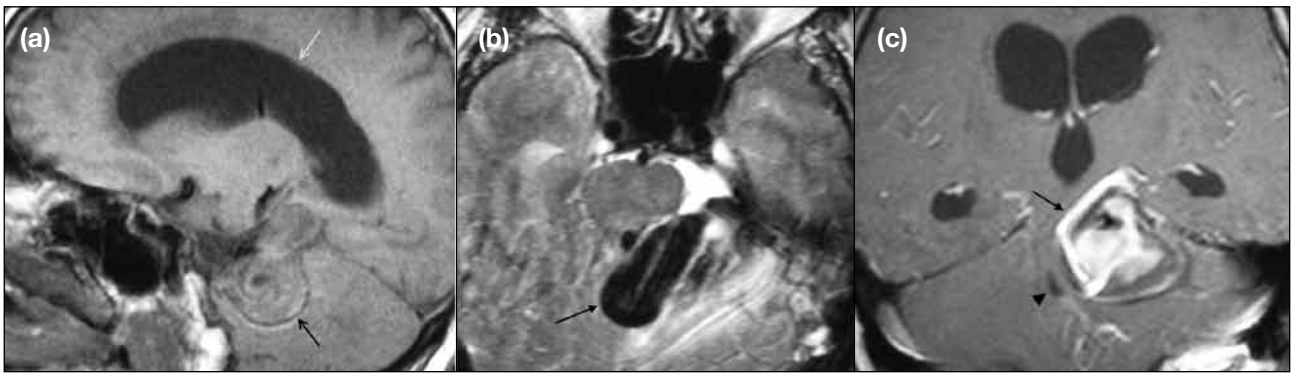

Figure 1. (a) There is a large heterogeneous signal lesion in keeping with a giant varix (black arrow) at the left cerebellopontine angle and quadrigeminal cistern on sagittal T1 sequence, causing obstructive hydrocephalus (white arrow), (b) which has a strong flow void (black arrow) on the axial T2 sequence. (c) Post-contrast coronal T1 sequence shows patchy contrast enhancement within the varix (black arrow). The lesion distorts the adjacent brainstem, cerebellum and the fourth ventricle (arrowhead) leading to obstructive dilatation of the lateral ventricles.

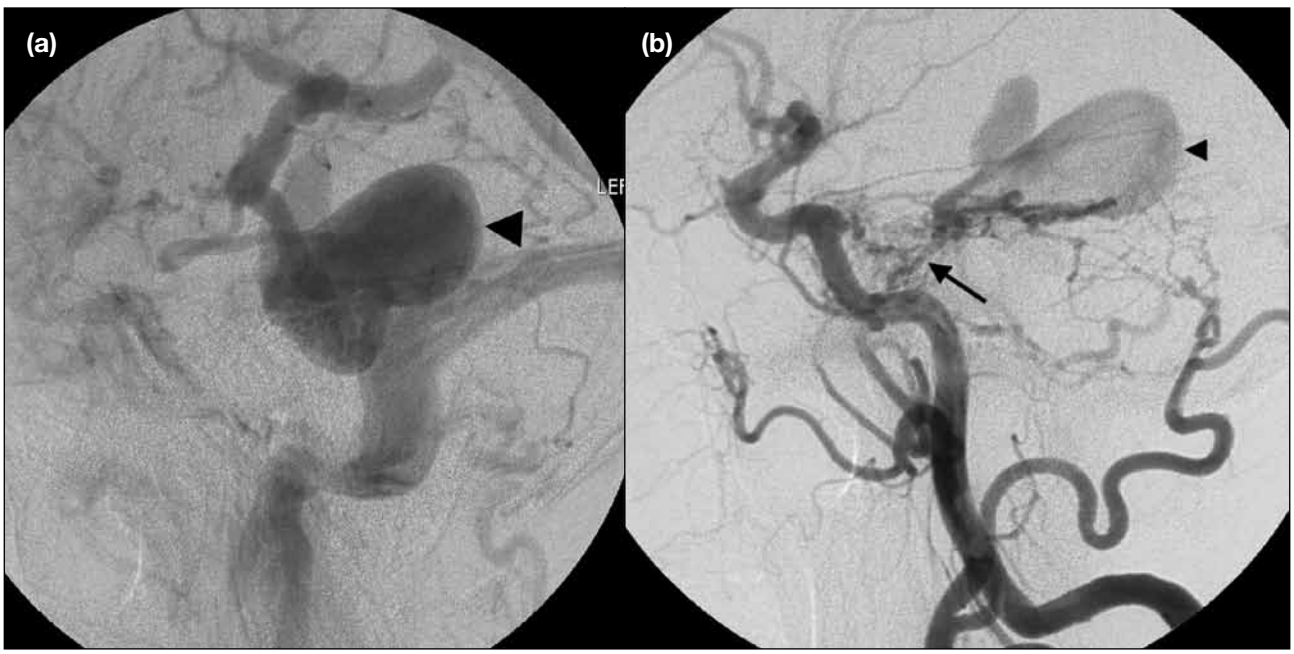

Figure 2. Selective catheter angiograms of the left internal carotid artery on (a) anteroposterior view and (b) oblique view, demonstrating a small arteriovenous fistula (arrow) arising from the small branches of the left distal internal carotid artery, which drained directly into the giant varix (arrowheads). The branches of the external carotid arteries were opacified as a result of the reflux of contrast from the internal carotid artery.

that time, as his symptoms resolved spontaneously over six weeks. There was no previous history of cerebral infections, neurosurgery, trauma, or malignancy. He re-presented five years later with a two-month history of headaches, ataxia, and left-sided hearing loss. Neurological examination revealed left ptosis and left facial sensory disturbances.

An MRI of the brain showed an irregular tubular mass measuring 35 mm in width and 45 mm in length in the left cerebellopontine angle and quadrigeminal cistern. The mass was compressing the fourth ventricle and cerebral aqueduct, causing hydrocephalus of the third and lateral ventricles (Figure 1). Hypointense T1 and T2 signals were found in the mass and thought to represent flow void in the mass lesion. Subsequent computed tomography angiography and catheter angiography demonstrated a left tentorial DAVF. It was fed by the meningohypophyseal trunk of the left distal internal carotid artery and the middle meningeal and occipital branches of the left external carotid artery. The DAVF drained into a giant varix (corresponding to the flow

void mass seen on MRI) in the left cerebellopontine angle and quadrigeminal cistern (Figure 2).

Endovascular embolisation was performed with the injection of 20% histoacryl and lipiodol into the middle meningeal and occipital branches of the left external carotid arteries. Polyvinyl alcohol particles were injected into the internal maxillary branches, which were the arterial feeders. A second-stage embolisation successfully obliterated the left meningohypophyseal trunk feeder with histoacryl diluted with lipiodol. The giant varix was successfully occluded after injecting two aliquots of 100% alcohol into the venous side of the fistula.

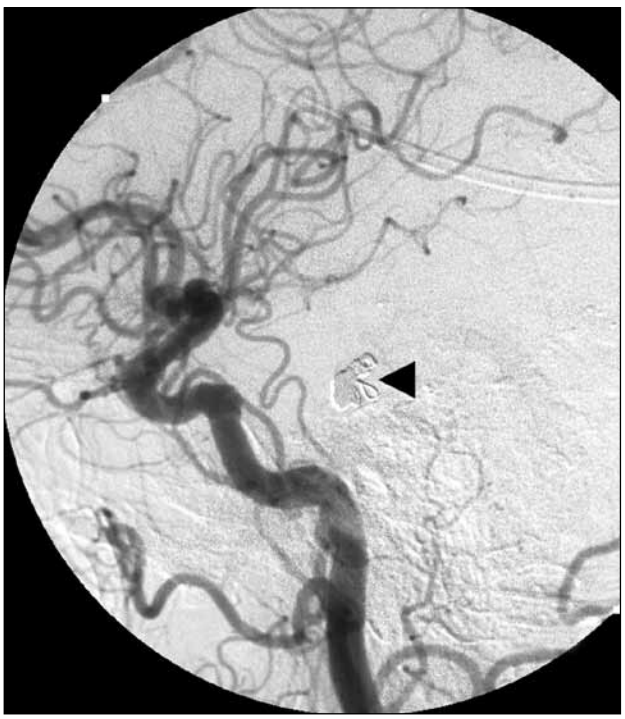

Figure 3. Follow-up catheter angiogram one month post-endovascular embolisation shows complete obliteration of the giant varix (arrowhead) and the tentorial dural arteriovenous fistula.

Follow-up catheter angiography one-month post-endovascular treatment showed complete obliteration of the giant varix and DAVF (Figure 3). The hydrocephalus was also successfully decompressed with a ventriculoperitoneal shunt insertion. Annual follow-ups with MRI consistently showed complete thrombosis of the varix and obliteration of the DAVF (Figure 4). The patient's symptoms also improved significantly with only mild residual ataxia.

DISCUSSION

DAVFs account for 10 to 15% of all intracranial arteriovenous lesions.[1] They are most commonly located at the transverse and cavernous sinus, with tentorial DAVFs (as in our case) being relatively rare and accounting for 2 to 9% of all cases.[2,3] On the other hand, varices were found in 11% of the 258 patients with intracranial DAVFs. The diameter of the varices reported in Cognard et al's

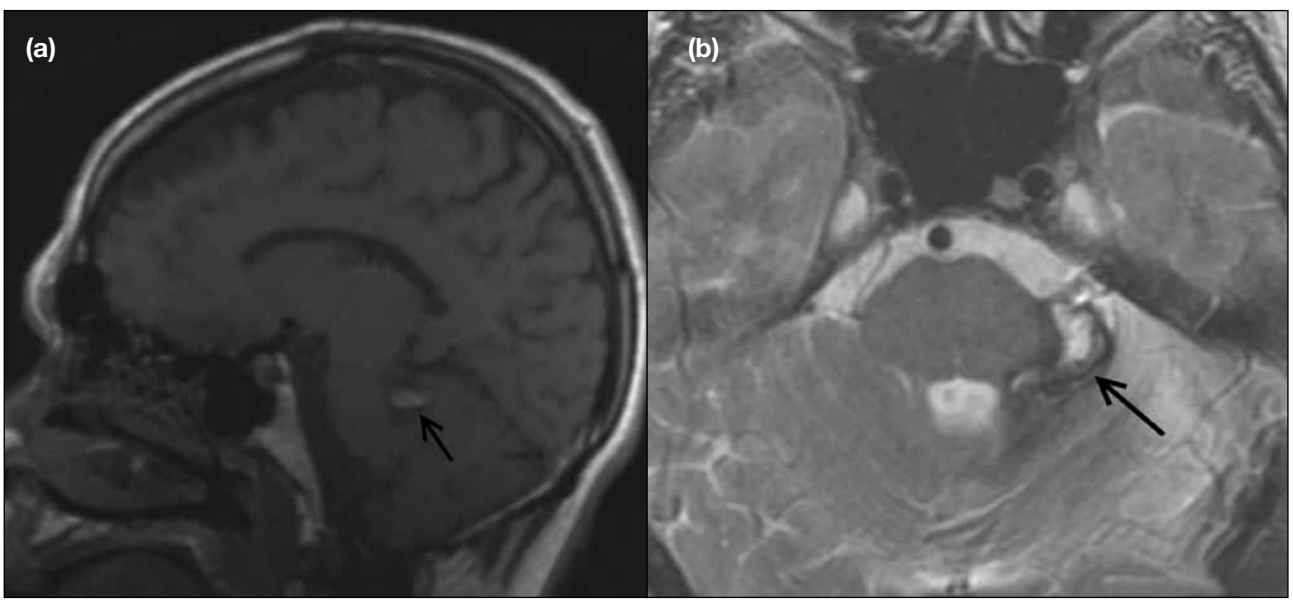

Figure 4. Follow-up magnetic resonance imaging three years post-endovascular embolisation shows marked reduction in the size of the varix (black arrows) with no significant mass effect on (a) the sagittal T1 sequence and (b) the axial T2 sequence. There is no flow void in the residual varix. The slight hyperintense signal on the T1 sequence and hyperintense signal on the T2 sequence within the varix are in keeping with thrombus.

study varied from 20 mm to 35 mm.[2] The presence of both conditions, namely the varix and tentorial DAVF, as found in our patient was reported in about 4% of cases,[2] which means they are extremely rare. Moreover, the size of the giant varix (35 mm x 45 mm) in our patient is larger than any reported in the previous literature.

It is postulated that the occlusion or thrombosis of the dural sinuses is the initial event that triggers the development of DAVFs.[4,5] Venous hypertension as a result of the thrombosis causes direct shunting between the arteries and veins, forming fistulae.[6] Suspected causes, including trauma, cerebral thrombophlebitis, infections, neurosurgery, hypercoagulable states, and pregnancy were identified in two series of patients with DAVFs.[2,3]

Classification

Cognard et al[2] had classified intracranial DAVFs into five categories according to the pattern of their venous drainage: type I — lesions were located in the main sinus with antegrade flow; type II — in the main sinus, with reflux into the sinus, cortical veins, or both; type III — with direct cortical venous drainage without venous ectasia; type IV — with direct venous drainage with venous ectasia; and type V — with spinal venous drainage. The venous drainage pattern and varix formation in our patient conformed to type IV, which is associated with a high risk of haemorrhage.[2]

Imaging Features of Intracranial Varices and Dural Arteriovenous Fistulae

An intracranial varix is described as venous ectasia, having a diameter of more than 5 mm and at least three-fold larger than that of the draining vein.[2] On the other hand, a DAVF is characterised angiographically by an immediate arteriovenous transition without a capillary bed or 'nidus' as occurs in an arteriovenous malformation. MRI may reveal a flow void cluster, adjacent brain parenchymal hyperintensity, a venous pouch, dilated vessels, vascular enhancement, and intracranial haemorrhage.[6] MR angiography can confirm the identifiable fistula, with venous flow–related enhancement and prominent extracranial vessels,[6] which were features well demonstrated in our case. In our patient, the unusual finding was a giant varix, but without an intracranial haemorrhage. The latter complication may occur in the presence of an intracranial varix or a tentorial DAVF, and often results in death.

Clinical Significance

Clinically, an intracranial varix can have a mass effect resulting in impaired drainage of the cerebrospinal fluid, leading to progressive intracranial hypertension and ventriculomegaly. Cognard et al[2] observed that 17% of their patients with type IV DAVFs developed aqueduct or third-ventricle obstruction secondary to the mass effect of a large venous ectasia. Similarly, our patient presented with neurological deficits and hydrocephalus resulting from brainstem compression due to the giant varix.

Generally, tentorial DAVF is associated with rapid neurological deterioration[2] and high risk of haemorrhage.[3,7] The risk of intracranial haemorrhage was reported to be about 66% in the presence of a varix associated with the intracranial DAVF and 58% in patients with a tentorial DAVF with or without varix.[2] Hence, the risk of haemorrhage in the presence of both conditions is extremely high. It is very unusual to encounter such a gigantic varix (as in our case) in the absence of any previous rupture.

Treatment

Due to the very high risk of intracranial haemorrhage and tendency to rapid neurological deterioration, prompt treatment of the giant varix and its associated tentorial DAVF is warranted.[2] This can be achieved by endovascular embolisation, surgery, radiosurgery, or a combination of these therapies. Combining endovascular and surgical interventions was previously reported as the optimal treatment for achieving complete obliteration of a tentorial DAVF.[3] Nevertheless, both the giant varix and the tentorial DAVF in our patient were successfully treated with endovascular embolisation alone.

In summary, simultaneous occurrence of a giant posterior fossa varix and tentorial DAVF is extremely rare and often associated with a high risk of life-threatening intracranial haemorrhage. Our patient's giant posterior fossa varix manifested itself as a mass effect causing hydrocephalus and compression on the brainstem, without any prior rupture. An understanding of the natural history and radiological characteristics of a simultaneously occurring posterior fossa varix and tentorial DAVF should prompt urgent diagnostic

and therapeutic initiatives to prevent fatal neurological sequelae.

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