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A treatable case of dementia—intracranial dural arteriovenous fistula

—個可治療的痴呆病例：顱內硬腦膜動靜脈瘻管

Dementia is a common medical problem that affects elderly patients. We report on a 77-year-old man with an intracranial dural arteriovenous fistula who presented with dementia that was initially thought to be irreversible and degenerative. Subsequent neuroendovascular intervention resulted in significant functional and cognitive improvement.

痴呆症是影響年長病人的常見醫療問題。我們報告一名77歲患有顱內硬腦膜動靜脈瘻管的男性，他出現痴呆症表徵，初期診斷認為無法復原，並會逐漸出現功能退化。在經過神經內血管治療後，機能和認知能力有重大的改善。

Introduction

Dementia is a clinical syndrome characterised by memory loss, intellectual decline, and personality change severe enough to interfere with daily functioning and the quality of life. According to a large-scale cohort study, the incidence of cognitive impairment for individuals aged over 70 years in Hong Kong is estimated to be 1.52% and 6.37% for men and women, respectively.[1] Most cases are due to Alzheimer's disease. Until recently, treatment has been confined to relief of symptoms with pharmacological therapy, such as cholinesterase inhibitors. Nonetheless every effort should be made to identify reversible and treatable causes of dementia. We present the case of an elderly man who had dementia as a result of a dural arteriovenous fistula (DAVF) that resulted in venous infarction of the brain. Trans-arterial embolisation was performed with consequent dramatic improvement in symptoms and brain-imaging abnormalities.

Case report

A 77-year-old man was admitted with a 1-week history of gradual deterioration in cognitive function. He had a previous right-sided infarct but no history of diabetes or hypertension. On examination he was found to have poor memory, slurring of speech, unsteady gait with frequent fall, and double incontinence. He was disorientated in time, place, and person, and his Glasgow Coma Scale (GCS) score was 14/15. He was un-cooperative during neurological examination. No cranial bruit could be detected and blood pressure and heart rate were within normal limits. Mini-Mental State Examination (MMSE) score was 0/30. Results of biochemistry and metabolic screening tests were unremarkable.

Initial unenhanced computed tomography (CT) scanning of the brain showed right parietal and multiple lacunar infarcts only with no hydrocephalus or space-occupying lesion such as meningioma, or chronic subdural haematoma. He was treated for infarct dementia with aspirin and followed up in the geriatric out-patient clinic.

Cognitive function continued to be poor and his behaviour became more aggressive. Repeated plain CT brain scan a year later revealed a heterogeneous lesion in the left parieto-occipital area with surrounding oedema. Subsequent brain CT with contrast showed a heterogeneous density in the left sigmoid sinus with dilated pial vessels suggestive of left sigmoid dural sinus thrombosis. Magnetic resonance imaging (MRI) of the brain and magnetic resonance venogram were arranged to confirm the diagnosis (Fig 1) and showed extensive

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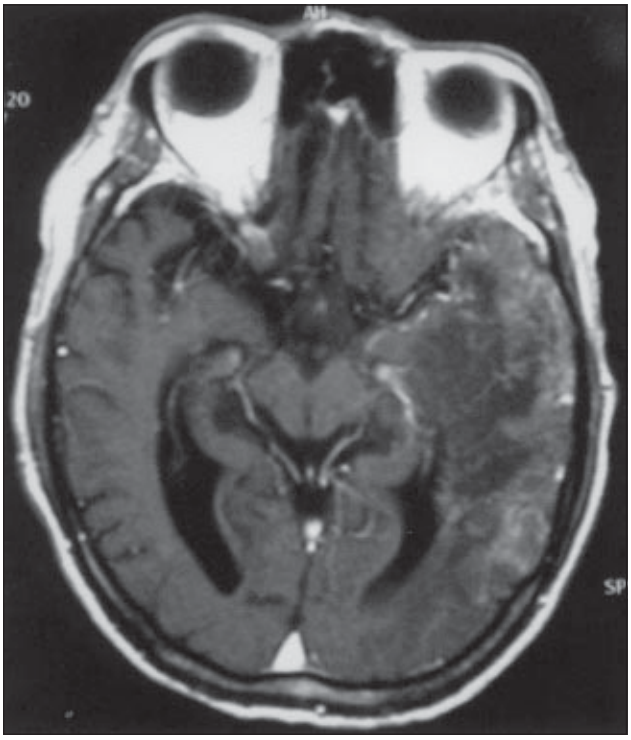

Fig 1. Cerebral magnetic resonance image showing extensive oedema involving left temporal and occipital lobes, extending to insular region

Intracerebral haemorrhage is evidenced. Left transverse sigmoid sinus is thrombosed with dilated pial and medullary veins suggestive of underlying dural arteriovenous fistula

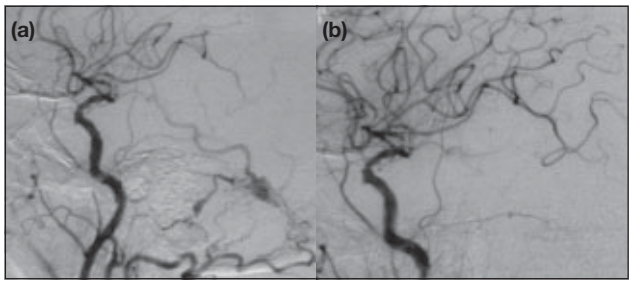

Fig 2. (a) Left vertebral angiogram (lateral view) showing a left transverse-sigmoid sinus dural arteriovenous fistula. It was supplied by left occipital and middle meningeal arteries and drained into vein of Labbe and other cortical veins. (b) Post-embolisation angiogram showing the dural arteriovenous fistula that was almost completely obliterated

oedema involving the left temporal and occipital lobes with intracerebral haemorrhage. There were dilated pial vessels in the sylvian fissure and cerebral hemisphere. The left transverse-sigmoid sinus was thrombosed; this may have explained the resulting venous infarction. These radiological features suggested the presence of a DAVF.

Cerebral angiogram (Fig 2) was performed under local anaesthesia and revealed that the DAVF was supplied by the left occipital and middle meningeal arteries and drained into the vein of Labbe and other cortical veins. The left occipital artery was embolised with polyvinyl alcohol particles and the left middle meningeal artery was embolised with Histoacryl-Lipiodol mixture. The fistula was almost completely obliterated. Post-procedure GCS score remained at 14/15 but the patient became more co-operative and less aggressive. Serial CT brain scan showed gradual resolution of oedema over the left temporal and occipital lobes. The MMSE score also improved from 0/30 before admission to 5/30. He gradually recovered and became ambulatory.

Discussion

Dural arteriovenous fistula is an abnormal connection between arteries and one or more veins or venous spaces called sinuses that are found in the covering of the brain. It accounts for 10% to 15% of all intracranial arteriovenous malformations.[2] The most common sites of involvement are the transverse, sigmoid, and cavernous sinuses.[3] Common causes of the DAVF include trauma, infections (eg mastoiditis), or thrombosis of the vein.[4-6]

Dural arteriovenous fistula is not a static lesion. It may behave aggressively and present with non-haemorrhagic neurological deficits or intracranial haemorrhage.[7] It can be classified according to the drainage vessels and the direction of flow and be benign or aggressive.[8] The aggressive type has retrograde cortical venous drainage that can cause venous hypertension and subsequently intracranial haemorrhage: the reported annual mortality varies widely but can be as high as 10.4% per year.[7] Spontaneous closure of DAVF is rare. Most spontaneously resolving lesions are type I DAVF.[9,10] To justify treating a cranial DAVF, the expected risk of sequelae during its natural course should be compared with the risk and the expected success rate of the proposed treatment.

Diagnosis may be difficult without imaging but DAVF should be suspected in patients who present with tinnitus, cranial bruit, and signs of raised intracranial pressure such as headache, blurred vision, bilateral papilloedema, and atrophic optic disc.[11,12]

Carotid duplex sonography of the external carotid artery can be used as an initial screening tool for diagnosis in patients with symptoms related to DAVF.[13] Computed tomographic and MRI scans may strongly suggest a DAVF in the posterior fossa, although they rarely are specific for such lesions.[14] In addition, CT brain scan can be normal in cases of fistula of the sinus without back flow into the cerebral vein. Angiography study, in addition to CT and MRI scanning, may aid in a more precise diagnosis of other cerebrovascular disorders, especially in younger or normotensive patients. The most typical feature on MRI or angiogram is a surplus of pial vessels with cortical venous

drainage. Another MRI finding of white matter oedema deep in the cerebral or cerebellar hemispheres is also direct evidence of venous congestion.[15,16]

Management options include close observation, neuroendovascular intervention, radiosurgery, and open surgery.[17] Embolisation is a safe and effective alternative to open surgery, and can be performed through a transarterial or transvenous route.[17,18] In our patient, transvenous embolisation was not possible because of thrombosis of the ipsilateral sigmoid sinus. Transarterial embolisation was performed with glue and resulted in near-complete obliteration of the DAVF. Transarterial embolisation is an effective treatment option although the fistula can recur. It can be retreated with second embolisation. Alternatively, open surgical excision of the fistula is technically feasible and more likely to be definitive with low morbidity. Stereotactic radiosurgery is another treatment option for DAVF, but complete obliteration of the DAVF takes time.[19] It should be reserved for benign DAVF when other treatment options have failed. Aggressive DAVF with retrograde cortical venous drainage requires urgent treatment with embolisation or open surgery.

Conclusion

The patient in this report presented with dementia initially thought to be untreatable. A high index of suspicion should be maintained in order to diagnose DAVF. The longstanding abnormalities as a result of venous hypertension and cerebral hypoperfusion are potentially reversible. Neuroendovascular intervention is a minimally invasive, effective, and reliable procedure that helps improve affective, cognitive, and behavioural abnormalities.

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