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Case Report

Dural arteriovenous fistula presenting as reversible dementia

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ABSTRACT

DAVF is an abnormal connection between arteries and veins, where supply is mainly through branches arising from the external carotid artery. A 30 years old male had multiple episodes of seizures and progressive behavioral and cognitive decline, inattention and disorientation to time, place and person, along with ptosis and ophthalmoplegia in the right eye. DSA provided a definitive diagnosis of hypervascular Dural AV fistula at the right transverse sigmoid junction with significant cortical venous hypertension. Through femoral access, both underwent transarterial embolization through the Middle Meningeal branch of the right external carotid artery using onyx, resulting in complete occlusion of fistula and resolution of venous hypertension. Post embolisation, cognitive function assessment revealed increased attention span and improved orientation with time, place and person. Thus, DAVF can be one of the rare but reversible causes of dementia. Early diagnosis and treatment may help to dramatically improve patients' clinical condition and minimize long-term disability.

Keywords: Dural AV fistula, Reversible dementia, Embolisation

INTRODUCTION

Most dementias such as Alzheimer's disease, Frontotemporal dementia are primarily neurodegenerative and irreversible. Dural AV fistula (DAVF) can be one of the rare but reversible causes, where early diagnosis and treatment may help to dramatically improve patients' clinical condition and minimize long-term disability.[1] The objective of this study was to recognize such causes of dementia that are potentially treatable and reversible.

CASE REPORT

30 years old male, farmer by occupation presented with history of two episodes of seizures with Right sided eye swelling and ptosis and progressive behavioral and cognitive changes since four months.

Four months ago, the patient had 2 episodes of generalised tonic clonic seizures. This was accompanied by progressive behavioral changes, where he became irritable and started talking irrelevantly.

It was accompanied by decreased food intake, refusal to take any medications and repeated bursts of anger on relatives. He stopped expressing any interest in family matters. He left his daily routine and stopped working on the farm. There were episodes of inappropriate laughing and crying. These symptoms progressed to the extent that he was not able to identify his family members and was totally dependent on them for his daily personal care activities. There was progressive right eye swelling, redness and incomplete ptosis.

On examination, his vitals were stable. He had severe cognitive impairment, inattention and disorientation to time, place and person. Forward attention span was 3 and backward was 0. MMSE was 8/30. FAB score was 3/18. Frontal release signs were present. Detailed higher function examination could not be performed due to severe

inattention. There was right sided eye severe congestion, ptosis and proptosis with complete external ophthalmoplegia. Remaining cranial nerves were normal. Motor examination was normal.

Patient underwent CT brain with angiogram was suggestive of white matter edema in bilateral high parietal region with dilated cortical veins, arterialization of Right transverse and superior sagittal sinus due to arteriovenous shunting, with enlarged right superior ophthalmic vein and bulging of right cavernous sinus.

Digital subtraction angiography provided a definitive diagnosis of hypervascular DAVF at the right transverse sigmoid junction with arterial feeders from right ECA and meningohypophyseal trunk of ICA, draining into the right transverse sinus. There was significant cortical venous hypertension noted (Figure 1).

Patient was planned for embolization of DAVF. Through a femoral access, he underwent transarterial embolization through middle meningeal branch of the right external carotid artery using onyx (18), resulting in complete occlusion of fistula and resolution of venous hypertension (Figure 2).

Patient was assessed on day 15 after embolisation. Cognitive function assessment revealed increased attention span with forward attention span of 5 and backward of 3 digits. He was oriented with time, place and person. MMSE was 16 /30. There was impaired judgement and abstract thinking. FAB score was 10/18.

Applause sign was positive but frontal release signs were absent with impaired graphical luria. Trail making tests were impaired (120 sec for trail A, could not complete trail B). His working and episodic memory was impaired but remote and semantic memory was intact. Language domain was normal. There was no apraxia and neglect. There was a significant decrease in right eye congestion, ptosis and proptosis.

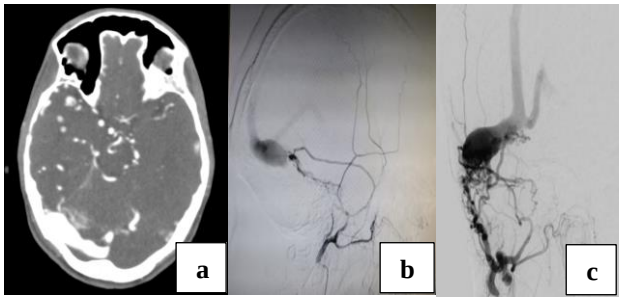

Figure 1: (a) Multiple abnormally dilated and tortuous vascular channels in the subarachnoid space; (b) enlarged right transverse sinus showing enhancement in the arterial phase s/o arterio-venous shunting; and (c) multiple abnormally dilated and tortuous vascular feeders connecting branches of the right ECA to the right transverse sinus.

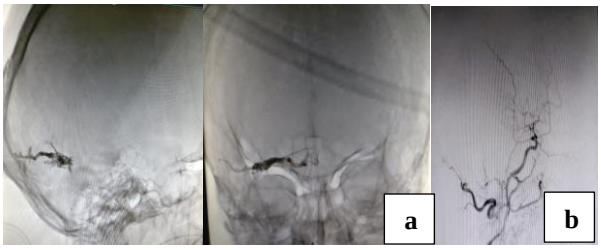

Figure 2: (a) Post right transverse sinus embolization showing glue cast; (b) right ECA shoot showing closure of the DAVF.

DISCUSSION

DAVF is an abnormal connection between branches of the external carotid artery, internal carotid and meningeal arteries and veins, which accounts for about 10% to 15% of vascular malformations in the brain.[2,3] The exact mechanism for development of DAVF is unknown but is believed to develop from acquired causes that cause increased pressure in the dural sinuses. The usual presentation is in the fifth or sixth decades of life.[4,5] The female-to-male ratio of 1:1.65.[2]

Patients with DAVFs may present with milder symptoms of orbital bruit, pulsatile tinnitus, headache, and ophthalmoplegia as well as the more severe presenting symptoms of neurological deficits and acute intracranial hemorrhage.[2,4] Rare presentations include DAVFs presenting with dementia presenting as a decline in neurocognitive function, and parkinsonism.[4,6]

The symptoms of DAVFs can also vary depending on location and pattern of cerebral venous drainage.[2,4,5] For example, the presentation of a fistulas draining into the cavernous sinus comprises of proptosis, visual loss, and ophthalmoplegia, whereas a DAVF of the transverse sinus may present with pulsatile tinnitus or parenchymal hemorrhage. Symptoms occur due to rise in the venous sinus pressure and volume, either via the DAVF or due to sinus obstruction due to thrombosis, resulting in harmful effects on brain parenchyma.[7]

Uncommonly, patients with DAVF may present with progressive dementia which occurs due to chronic hypoperfusion. There may be 2 possible mechanisms of DAVF-induced dementia namely, cortical dementia and thalamic dementia.[8] In cortical variant, the mechanism appears to be CSF disturbance leading to medullary venous congestion secondary to occlusion or venous hypertension resulting in arterialization of medullary veins as a result of direct reflux into a medullary vein from the DAVF, causing parenchymal edema and impaired function of affected regions which has been described as congestive venous encephalopathy. Impaired CSF resorption can also lead to secondary hydrocephalus. Venous congestion and ischemia of thalamus due to impaired deep venous drainage is responsible for thalamic dementia. There may be tentorial venous reflux to straight

sinus and vein of Galen and from there into internal cerebral veins and basal veins. Decreased frontal lobe perfusion may lead to symptoms such as decreased speech output. Seizures that occur may be more likely from venous congestion than from hemorrhage.

Contrast-enhanced CT imaging may show tortuous feeding arteries and dilated cortical draining veins. MRI T1/T2WI flow voids may show thrombosed sinus. Digital subtraction angiography (DSA) reveals predominance of vascular supply from meningeal arteries, cortical veins and retrograde venous drainage. Endovascular approaches are the treatment of choice in most DAVFs.[9] Various approaches (transarterial, transvenous, combined transarterial/ transvenous, or transorbital approach) can be used to cure a DAVF endovascularly depending on anatomy. Liquid embolic agents and coils are usually used as primary materials.[10] In refractory cases, a combined endovascular and neurosurgical approach is recommended.[11]

CONCLUSION

DAVF can be one of the rare but reversible causes of dementia, where early diagnosis and treatment may help to dramatically improve patients' clinical condition and minimize long-term disability. There have been major advancements in the endovascular treatment of DAVF, with improved success rates of procedures.

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