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## Dural arteriovenous fistulas with aggressive course: clinical and angiographic correlations in two patients

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**Abstract** Cranial dural arteriovenous fistulas (DAVFs) usually present with non-aggressive symptoms. We here report two patients who presented a peculiar clinical picture related to DAVFs, with focal neurological signs and haemorrhagic (case 1) or ischaemic lesions (case 2) respectively. The clinical and angiographic findings and putative pathophysiological mechanisms are discussed.

**Key words** Stroke • Dural arteriovenous fistula • Cerebral angiography

### Introduction

Cranial dural arteriovenous fistulas (DAVFs) constitute 10%–15% of intracranial vascular malformations [1–3]. They usually present with non-aggressive symptoms such as isolated headache, bruit, and ocular symptoms not related to intracranial hypertension [1, 3]. Sometimes, patients present with aggressive symptoms, including intracranial hypertension, seizures and, less frequently, stroke [1, 3–5]. Cerebral angiography remains the gold standard for the diagnosis of DAVFs. We here report the clinical and angiographic picture of two patients with DAVFs, both exhibiting peculiar features, with aggressive symptoms.

### Case report

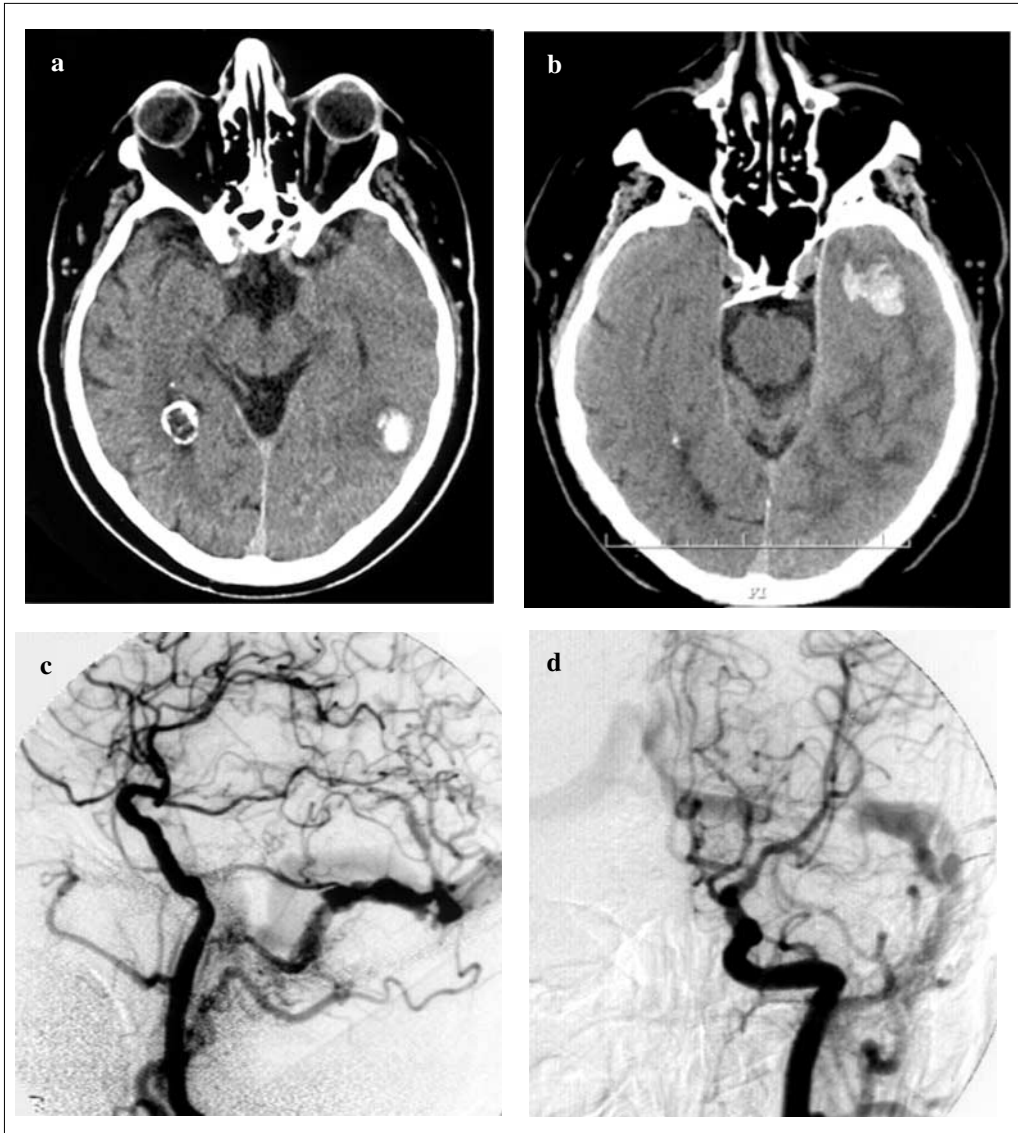
#### Case 1

An 80-year-old man, with a history of hypertension pharmacologically treated, was admitted to our institute for global aphasia and headache of sudden onset. A brain CT scan was immediately performed and showed a haemorrhagic lesion, 2 cm in size, located in the posterior part of the left temporal lobe (Fig. 1a). Blood parameters, including coagulation factors, were normal. In the following days, the clinical picture progressively improved. Before discharge, the neurological evaluation showed a mild comprehensive aphasia and a new CT scan disclosed a significant reduction of the haemorrhagic lesion. One month later the patient was admitted again for headache and worsening of aphasia. The neurological examination evidenced the presence of global aphasia and a CT scan showed a new haemorrhagic lesion in the left temporal lobe, located more anteriorly than the previous one (Fig. 1b). At that time the patient underwent an angiographic procedure, which showed the presence of a dural fistula, fed by the left occipital artery and other branches (the ascending pha-

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**Fig. 1a-d** *Neuroradiological findings in case 1. a* CT scan performed after the first episode of aphasia, showing a small left posterior temporal haemorrhage; *b* CT scan 1 month later, showing a new haemorrhage in the anterior temporal lobe; *c,d* angiography showing a fistula of the left transverse sinus, fed by an ectatic occipital artery, and other branches of the external carotid artery (the ascending pharyngeal and posterior auricular arteries) with partial occlusion of the sinus and retrograde drainage through cortical veins of the temporal lobe

ryngeal and posterior auricular arteries) of the external carotid artery, involving the left transverse sinus (which was partially occluded), with abnormal recruitment of cortical veins (Fig. 1c,d). The patient refused therapeutical embolisation of the DAVF.

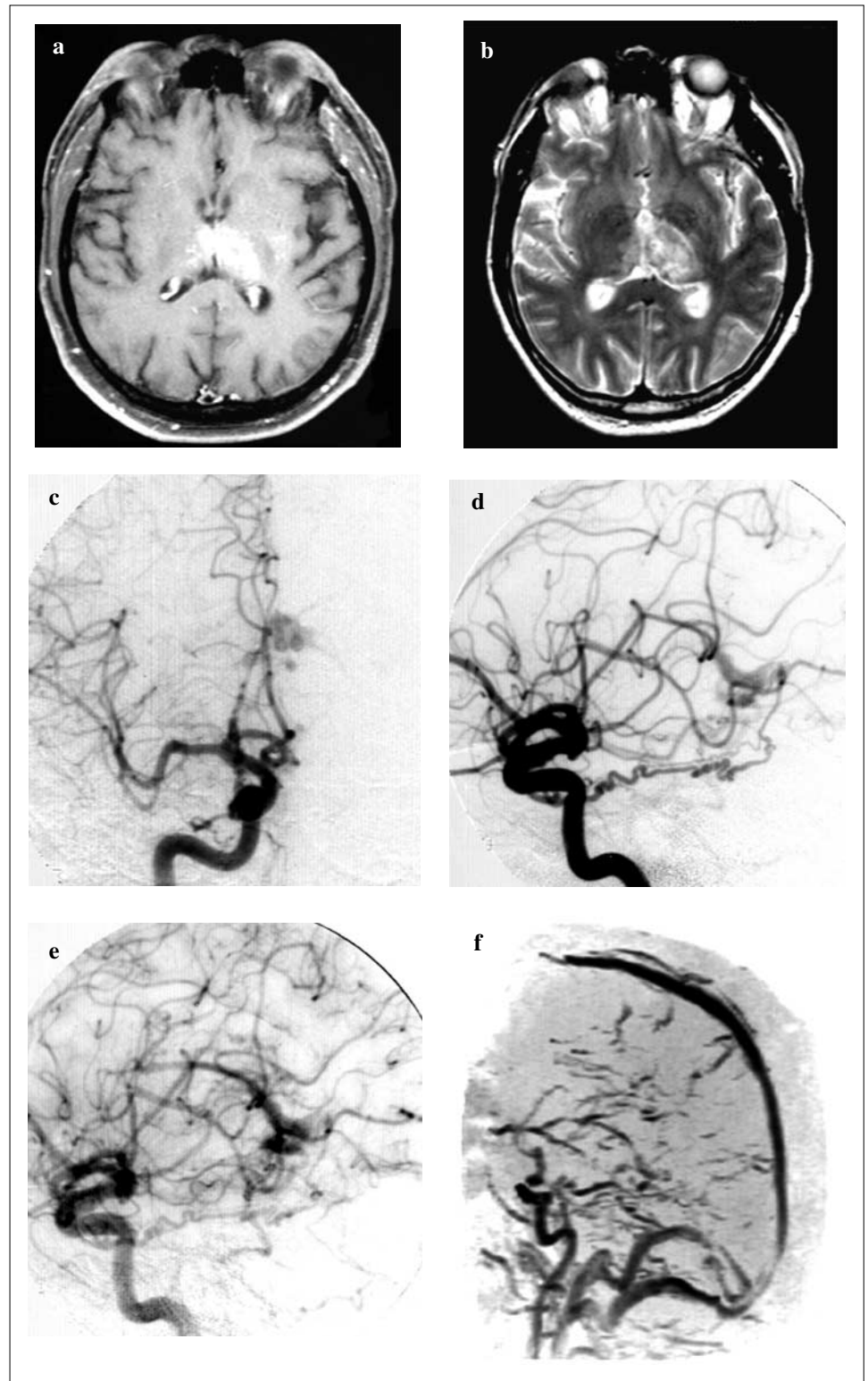
## Case 2

A 65-year-old man was admitted to our institute because he suddenly became confused and disoriented as to time and space. A CT scan performed a few hours after admission was normal. A cerebral magnetic resonance imaging (MRI) performed 3 days later showed the presence of a large ischaemic lesion located bilaterally in the posterior thalamus (mainly on the left side) (Fig. 2a). The neurological examination showed a global amnesic deficit, as confirmed by neuropsy-

chological tests (Mini Mental Examination, Milan Overall Dementia). No coagulation disorders were detected through extensive laboratory evaluation. The angio-MRI failed to inject the straight sinus (Fig. 2f); the cerebral angiography (15 days later) showed a DAVF between both tentorial arteries (mainly the right) and the right internal cerebral vein (Fig. 2c-e). The patient declined the fistula embolisation procedure. At discharge, the neurological examination showed a mild improvement of the amnesic syndrome, which was still evident one year later.

## Discussion

DAVFs are characterised by the presence of an abnormal arteriovenous fistula located in the dura. The natural course of intracranial DAVFs is generally benign. However, some



**Fig. 2a-f** Neuro-radiological findings in case 2. **a** contrast MRI performed 3 days after the onset of amnesic syndrome showing ischaemic changes bilaterally in the thalamus (prevalent on the left); **b** MRI 15 days later confirming a bilateral thalamic lesion (prevalent on the left); **c,d** right-sided carotid angiography (15 days later) showing an ectatic tentorial artery and early filling of the vein of Galen; **e** retrograde filling of the right internal cerebral vein with no filling of the straight sinus; **f** Angio-MRI showing no injection of the straight sinus

patients may present aggressive neurological symptoms [1, 4, 5]. Among aggressive symptoms, haemorrhage is relatively uncommon and ischaemia very rare [1–3, 6, 7]. The two patients reported here presented a peculiar clinical picture related to DAVFs with focal neurological signs associ-

ated with haemorrhagic (case 1) and ischaemic lesions (case 2).

It has been suggested that the high variability of symptoms in DAVFs may depend on different factors. More specifically, it has been reported [1, 3] that lesions drain-

ing into a dural sinus produce only benign symptoms, whereas cortical venous drainage is frequently associated with aggressive neurological symptoms. Awad et al. [6] reported that an aggressive presentation often occurs with tentorial and anterior cranial fossa fistulas, probably because of their cortical venous drainage. More recently Borden et al. [2] suggested a simplified classification into three categories: Type I, with a supply via meningeal arteries and anterograde drainage into venous sinus; Type II, supply via meningeal arteries, anterograde or retrograde drainage into dural venous sinus/meningeal vein and retrograde leptomeningeal venous drainage; Type III, supply via meningeal arteries, retrograde leptomeningeal venous drainage. According to this classification, Types II and III frequently have an aggressive presentation. All of these findings suggest that the variability of symptoms in DAVFs depends mainly on the morphology of the fistula drainage and only partially on their location [1, 8, 9].

Among dural fistulas, the fistula of the transverse sinus is the most frequent and usually presents with non-aggressive symptoms [1], because it generally drains into the sinus only. However, case 1 harbouring a fistula of the transverse sinus presented an aggressive behaviour, with repeated intracerebral bleeding. It has been proposed [1] that the different types of venous drainage correspond to different stages in the course of the disease, and Borden et al. [2] reported a progression of dural fistulas from one type to another. Indeed, a partial sinus thrombosis can redirect arterialized blood into a vein, and a progressive sinus thrombosis can transform one type of DAVF into another, with a switch from benign to aggressive symptoms. In patient 1, the progression of the transverse sinus thrombosis may constitute the main factor leading to increased risk of haemorrhage.

Contrary to the transverse sinus fistulas, tentorial fistulas often present with aggressive symptoms [8, 9]. Cognard et al. [1] reported that all dural fistulas in the tentorium have a cortical venous drainage, with the most frequent disclosure being the haemorrhage [3, 10]. Patient 2 is quite peculiar for two reasons: first because he presented with an aggressive symptomatology such as an amnesic-confusional syndrome, and second because these symptoms were due to a bilateral ischaemia of the posterior part of the thalamus. Cognitive impairment associated with DAVFs is an uncommon feature [1, 6, 10]. More recently, Hurst et al. [10] reported that 5 out of 40 reviewed patients with DAVFs presented progressive cognitive changes caused by an impairment of cerebral venous drainage. The authors suggest that the result is a widespread venous hypertension causing diffuse ischaemia and progressive dysfunction of brain parenchyma. In this regard, the acute onset of cognitive impairment in our patient associated with a focal ischaemic lesion of the thalamus constitutes a rare variant in the presentation of DAVFs, and – to our knowledge – it has never been reported. It is conceivable that the presence of the fistula with the tentorial arteries draining to the

straight sinus primarily and retrogradely to the internal cerebral veins has determined progressive thrombosis of the straight sinus, with consequent retrograde venous congestion [8]. The final occlusion of the straight sinus has led to significant haemodynamic derangement, with sudden increase in venous congestion and development of ischaemia in the territory drained by the internal cerebral veins. In the literature another case of bilateral thalamic infarction in the presence of an arteria-venous fistula between the anterior choroidal artery and the deep venous system has been reported [11], and the genesis of the infarction attributed to the engorgement of the venous system. The responsibility of venous hypertension in the aggressive symptomatology of many DAVFs has been already stressed by other authors [6, 8]. The hypothesis that the DAVFs observed in our patient are a secondary expression of venous thrombosis should be discarded, as the formation of a fistula determined by occlusion of a sinus is a phenomenon possibly taking months or years to occur, and not a few days, as observed in our case.

The present study demonstrates that an ischaemic stroke, in addition to a haemorrhagic event, may cause an aggressive clinical course of DAVFs. Therefore, the possibility of a DAVF-related stroke should be considered in the diagnostic protocol for cerebrovascular diseases, also in elder patients.

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