
CASE REPORT

Carotid-cavernous Sinus Fistula Presenting with Contralateral Intracerebral Haemorrhage

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ABSTRACT

Carotid-cavernous fistula is a well-established neurological disease that typically presents with proptosis, chemosis, ophthalmic bruit, and cranial nerve palsies. This report is of an atypical case of carotid-cavernous fistula that resulted in contralateral intracerebral haemorrhage. A 28-year-old woman presented with sudden headache and was found to have a right temporal intracerebral haemorrhage. She did not have ocular symptoms such as proptosis or chemosis. Angiographic studies revealed a left-sided direct-type (Barrow type A) carotid-cavernous fistula and no other significant findings. The left type A carotid-cavernous fistula was likely to have caused venous hypertension around the contralateral cerebral hemisphere and intracerebral haemorrhage by means of intercavernous venous cross-flow. This patient was also unusual in that the type A carotid-cavernous fistula, which rarely resolves without intervention, had disappeared spontaneously four days later. Carotid-cavernous fistula should be considered as a differential diagnosis in patients with spontaneous intracerebral haemorrhage, even in the absence of ocular symptoms. Investigations should include thorough angiographic studies of the contralateral cerebral vessels.

Key Words: Angiography; Carotid artery diseases; Cavernous sinus; Cerebral hemorrhage; Stroke

中文摘要

頸內動脈海綿竇瘻引致的對側腦內出血

劉泳恆、梁嘉傑、李雷釗、呂偉文

頸內動脈海綿竇瘻是一種已知的神經性疾病，症狀為眼球突出、球結膜水腫、眼雜音及顱神經麻痺。本文報告一宗頸內動脈海綿竇瘻的非典型病例，病徵為對側腦內出血。一名28歲女性有突發性頭痛，發現為右腦顳叶出血。病人未有出現如眼球突出或球結膜水腫，但其血管造影結果顯示左側一直接型（即甲型Barrow）的頸內動脈海綿竇瘻。除此以外，並無其他嚴重疾病。此左邊甲型海綿竇瘻造成竇內靜脈對流，引致對側大腦半球的靜脈高壓和腦內出血。這種甲型海綿竇瘻很少會自行緩解。可是這名病人的頸內動脈海綿竇瘻在四日後自動緩解。如果病人出現自發性腦出血，縱使未有眼部症狀，都應考慮頸內動脈海綿竇瘻鑒別診斷的可能性。應為病人的對側腦血管進行徹底的血管造影檢查。

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INTRODUCTION

Carotid-cavernous fistula (CCF) is an abnormal communication between the venous channels and the internal carotid artery (ICA) within the cavernous sinus. The clinical presentations are characterised by the onset of venous hypertension within and around the cavernous sinus. Patients typically present with chemosis, pulsatile exophthalmos, cranial nerve palsies, diplopia, visual loss, and ophthalmic bruit as a result of engorgement of the ophthalmic veins.¹⁻³ Less commonly, retrograde venous reflux into the superficial cerebral venous system may also result in intracerebral haemorrhage (ICH) in the adjacent fronto-temporal lobes.⁴⁻⁶ We report an unusual case of CCF in a woman who presented with contralateral ICH without ocular symptoms.

CASE REPORT

A 28-year-old woman with good past health presented in December 2005 with sudden-onset severe headache. There was no history of trauma. Neurological examination was normal. Computed tomography (CT) demonstrated a right temporal lobe ICH (Figure 1). Digital subtraction angiography was subsequently performed to look for the underlying cause, and revealed the presence of a CCF supplied by the intracavernous left ICA (Figure 2) with venous cross-flow between the left and the right cavernous sinus. The right ICA and left external carotid artery (ECA) angiograms were unremarkable, with no evidence of meningeal supply to

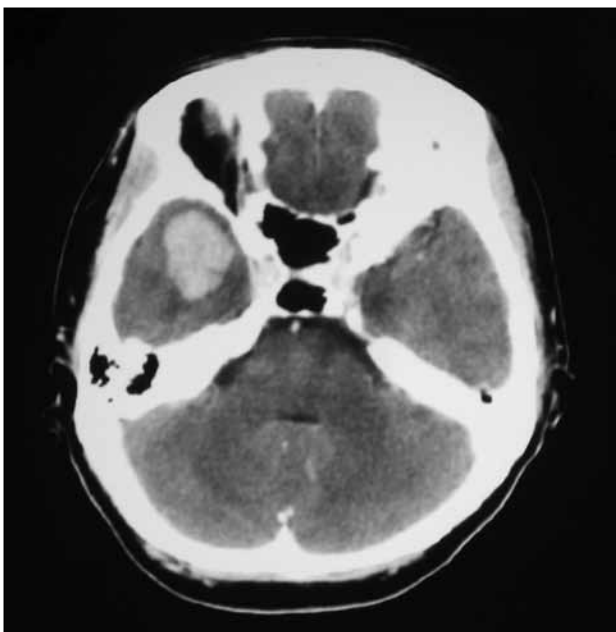


Figure 1. An axial non-contrast computed tomography image showing a right temporal intraparenchymal haemorrhage.

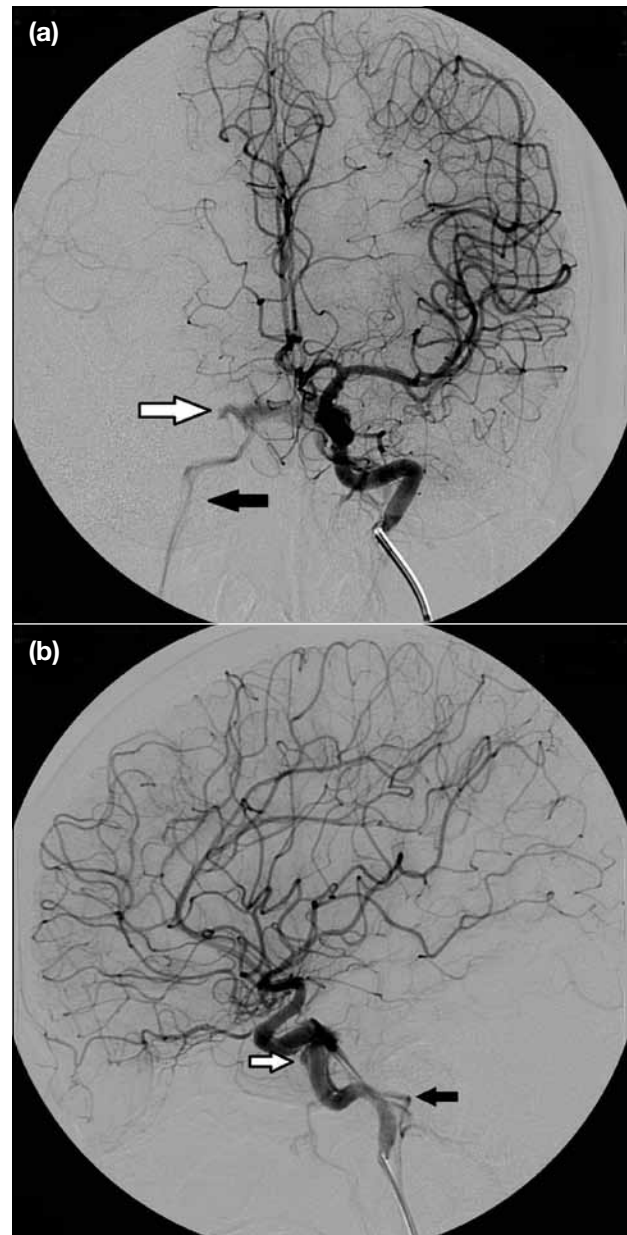


Figure 2. (a) Anteroposterior and (b) lateral digital subtraction angiograms showing a direct-type (type A) carotid-cavernous fistula supplied by the left internal carotid artery. There was cross-flow to the contralateral cavernous sinus with opacification of the contralateral sphenoparietal sinus (white arrows), possibly causing increased venous pressure and intraparenchymal haemorrhage. Drainage was via the inferior petrosal sinuses (black arrows). Note that the superior ophthalmic veins were not dilated.

the CCF. No carotid artery aneurysm was found. The presumptive diagnosis was left ICA CCF (Barrow type A) causing venous hypertension through cross-flow resulting in spontaneous contralateral ICH.

Endovascular embolisation of the CCF was planned and repeat angiography was performed four days later.

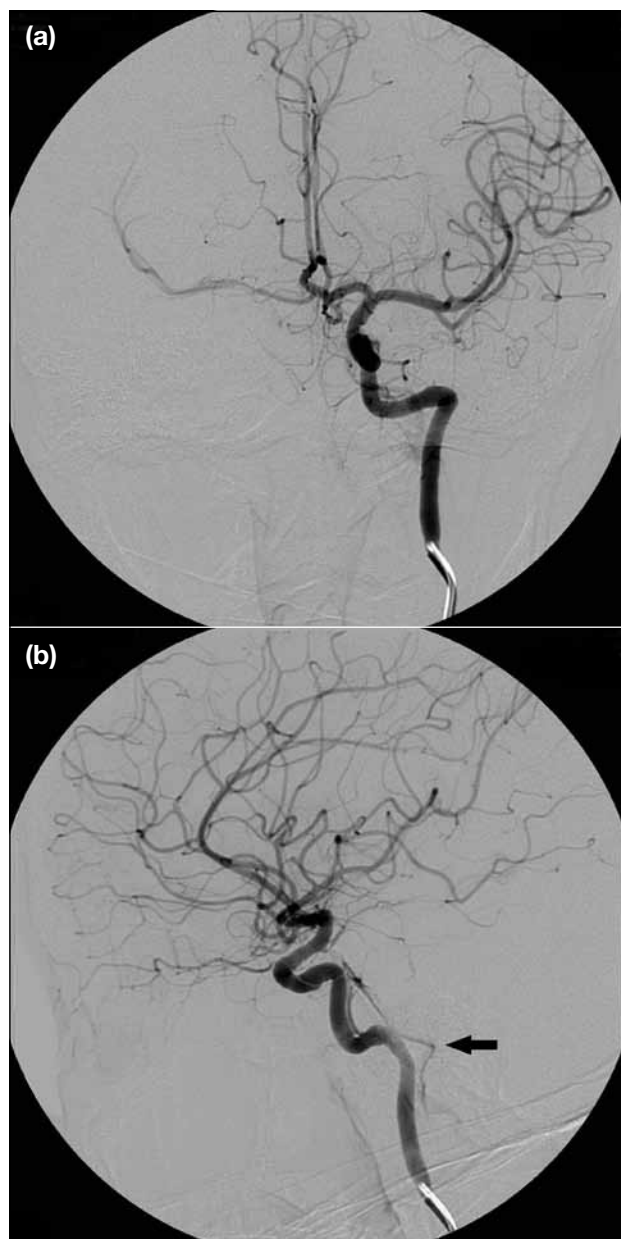


Figure 3. (a) Anteroposterior and (b) lateral digital subtraction angiograms performed after four days showing spontaneous closure of the carotid-cavernous fistula. Minute opacification of the left cavernous sinus with drainage via the left inferior petrosal sinus (black arrow) into the left internal jugular vein was noted.

Left ICA angiogram showed only minute opacification of the left cavernous sinus without further cross-flow to the right side, and the CCF had resolved spontaneously (Figure 3). Therefore, no embolisation was carried out.

Clinically, the patient's headache showed gradual resolution. Repeat CT of the brain performed three months later showed complete resolution of the right temporal ICH.

DISCUSSION

CCF can be classified based on the anatomy, pathophysiology, and aetiology. One of the most widely accepted anatomic classifications is Barrow's classification¹ – type A is defined by direct high-flow shunts between the ICA and the cavernous sinus; type B is defined by dural shunts between the meningeal branches of the ICA and the cavernous sinus; type C is defined by dural shunts between the meningeal branches of the ECA and the cavernous sinus; and type D is defined by dural shunts between the cavernous sinus and meningeal branches of both the ICA and ECA. This patient was therefore classified as having type A CCF.

Type A CCF most commonly results from cranial trauma, iatrogenic injury, rupture of an intercavernous carotid artery aneurysm, or in association with disorders of collagen deposition.² In this patient, however, no discernable predisposing cause was found. The typical presentation of ocular symptoms and angiographic evidence of ophthalmic vein engorgement were absent. Instead, significant intercavernous venous cross-flow had resulted in venous hypertension in the right-sided cavernous sinus and sphenoparietal sinus. Subsequent venous hypertension in the superficial middle cerebral veins, which typically empty into the cavernous sinus and sphenoparietal sinus, resulted in this unusual presentation of a contralateral ICH.^{5,6} To the authors' knowledge, there have been no previous reports of spontaneous ICH secondary to a contralateral CCF.^{4,7,8}

This case was also unusual because of the spontaneous resolution of the CCF. The management of CCF varies with the type of shunting found. While 43 to 62% of indirect (types B, C, and D) CCF may resolve without intervention, spontaneous resolution is distinctly uncommon for the direct-type (type A) CCF.^{5,9} Goto et al⁸ reported on 148 patients with type A fistulae. Only 3% of the CCFs had spontaneous resolution, and 2.7% of the patients had died from haemorrhagic complications. Early and aggressive treatment is therefore indicated for a type A fistula.³

Currently, patients, especially in the younger age-group, who present with ICH undergo CT or magnetic resonance imaging (MRI) before proceeding to angiography. MRI features of CCF include abnormal flow voids in the cavernous sinus and complications related to venous hypertension such as an enlarged cavernous sinus, engorged draining veins, and swollen extra-ocular muscles.^{10,11} The source images of three-

dimensional time-of-flight magnetic resonance angiography are particularly useful in making the diagnosis.¹² A study has also shown the effectiveness of CT angiography in showing an enlarged enhancing cavernous sinus or draining veins, although conventional angiography is still the gold standard in making the diagnosis.¹³

CONCLUSION

In direct-type (type A) CCF, the intercavernous venous cross-flow may result in ICH in the contralateral hemisphere. Spontaneous resolution is possible, as seen in this patient. In young patients with spontaneous ICH, for which angiography is usually employed in an investigation for underlying aneurysm or vascular malformation, CCF should be considered a differential diagnosis and contralateral angiogram should also be performed.

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