

Brainstem Hemorrhage Caused by Direct Carotid-Cavernous Fistula

A Case Report and Literature Review

FOOK-HOW CHAN^{1,4}, CHAO-YU SHEN^{2,3,4}, JUNG-TUNG LIU^{1,4}, CHO-SHUN LI^{1,4}

¹ Department of Neurosurgery, ² Department of Medical Imaging, ³ School of Medical Imaging and Radiological Sciences, ⁴ School of Medicine, Chung Shan Medical University Hospital; Taichung, Taiwan

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Summary

A 34-year-old woman presented with a history of persisting headache for years, and a newly developed dizziness, left facial palsy and right hemiparesis two days prior to this admission. Initial computed tomographic angiography of the head demonstrated an area of increased density in the left middle and posterior fossae. Multiple aneurysmally dilated venous ectasias with contrast enhancement at the left pre-pontine cistern causing a massive mass effect to the brainstem were also noted, suggesting a huge vascular abnormality. Digital subtraction angiography revealed an abnormal vascular lesion surrounding the brainstem, which indicated a left direct carotid-cavernous fistula with posterior drainage. As her consciousness deteriorated the next day, a follow-up computed tomography scan was done which revealed a pontine hemorrhage. Subsequently, endovascular closure of the fistula with sacrifice of the left ICA was performed, which successfully eliminated the imaging abnormalities.

Introduction

Carotid-cavernous fistula (CCF) is an acquired lesion caused by arteriovenous shunts between the high-pressure carotid artery and low-pressure venous system of the cavernous sinus. The signs and symptoms of CCF result from the congestive venous status around the cavernous sinus. The typical clinical signs include the triad of pulsatile exophthalmos, prop-

osis, bruit and chemosis. Other atypical signs including cranial nerve palsy, hemiparesis and intracranial bleeding have also been reported, of which, brainstem dysfunction is an extremely rare complication caused by CCF.

Case Report

A 34-year-old woman was admitted to our neurosurgical service because of progressive headache, dizziness, left facial palsy and right hemiparesis for two days. Initial clinical examination revealed a bruit in the left orbit but there was no chemosis or exophthalmos. Her consciousness was clear. Motor examination revealed her muscle power was weaker on the right limbs as 3/5 (on The British Medical Research Council strength scale) with a mild increase in deep tendon reflex. Ocular examination showed a dilated pupil, complete loss of ocular movements and visual acuity in her left eye. Computed tomographic angiography (CTA) of the head demonstrated an area of increased density in the left middle and posterior fossae with filling of the left superior ophthalmic vein. Multiple venous ectasias with contrast enhancement at the left pre-pontine cistern causing a massive mass effect to the brainstem were also noted, suggesting a huge vascular abnormality (Figure 1). A symptomatic venous pouch was discovered with partial thrombosis, and a fluid-fluid level was found in the wall of the venous pouch.

Tracing back the history, it emerged that the patient had been involved in a severe motor-

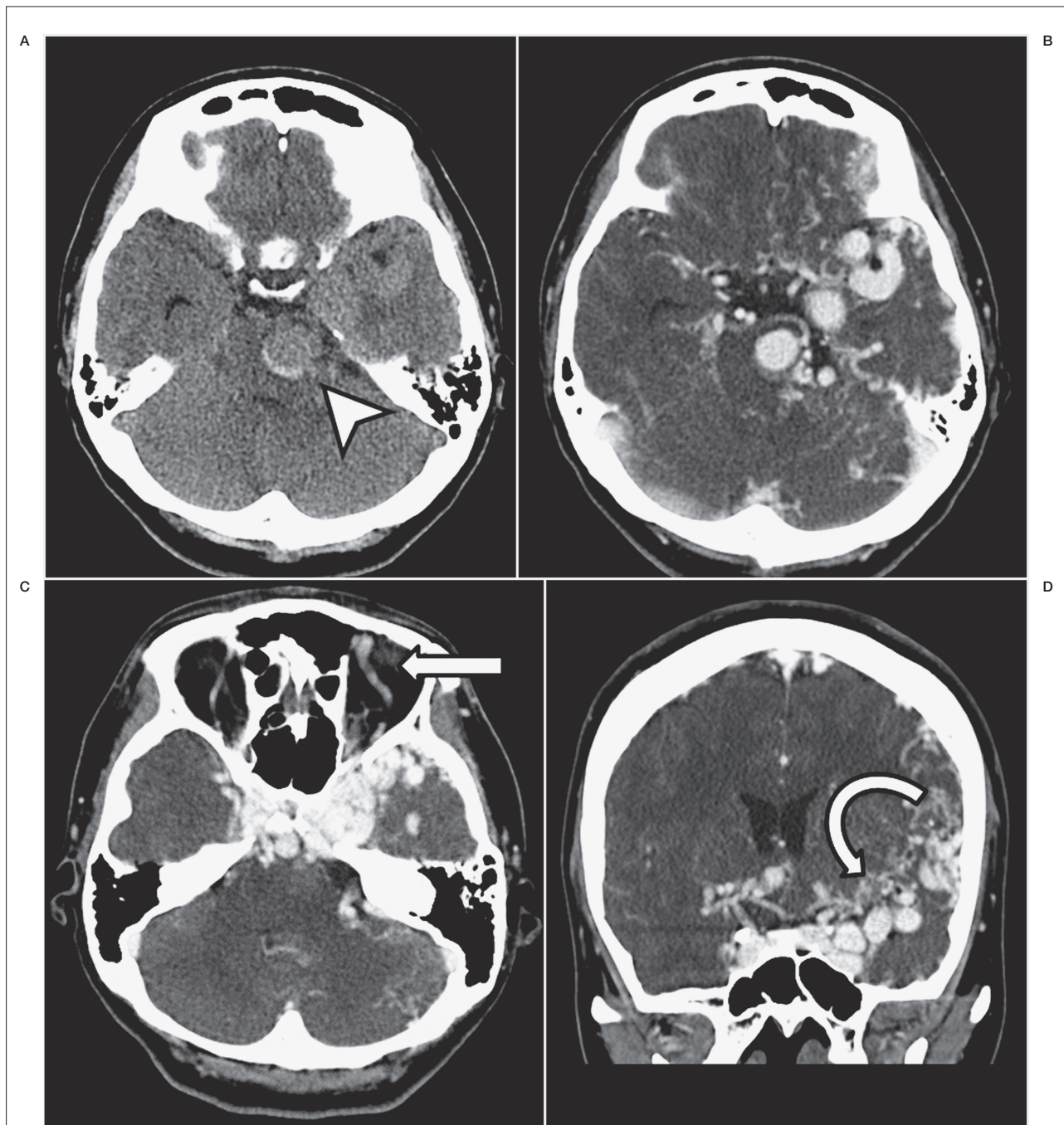


Figure 1 A) CT scan shows an aneurysmally dilated venous pouch with wall thrombosis compressing the left pons (arrow-head). CT angiography shows the characteristic prominence of the left ophthalmic vein (C, straight arrow) and multiple enhancing, engorged vessels in the left cerebrum, cerebellum and brainstem (D, curved arrow).

bike accident seven years earlier, resulting in a complete loss of visual acuity in the left eye and a remarkable fracture of the left zygomaticomaxillary complex and maxilla extending to the right submandibular bone. Since the accident, she had experienced chronic headache, sleeping disorder and major depression, and

received treatment at the outpatient department.

Digital subtraction angiography (DSA) was also performed, confirming a left high-flow direct CCF consistent with Barrow type A. This direct CCF presented a complete steal phenomenon from the left internal carotid artery

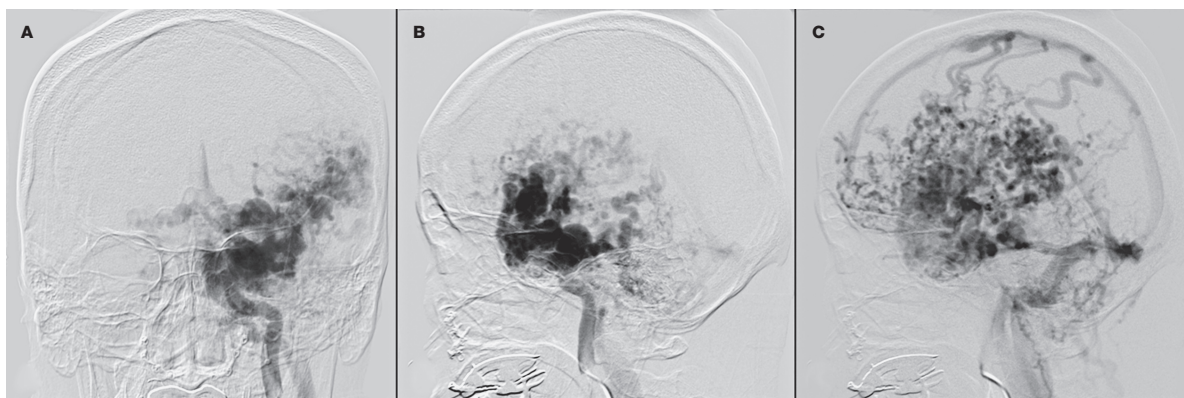


Figure 2 A-E) DSA of the left ICA confirms a direct, high flow left CCF. A) The anteroposterior projection of the arterial phase of the left ICA demonstrates a complete steal phenomenon from the left ICA into the left cavernous sinus. The lateral projections of early arterial phases (B) and venous phases (C) of the left ICA show a left direct CCF with retrograde flow into both superficial and deep venous system, causing marked tortuosity of the affected venous vessels with multiple focal dilated venous ectasias. The anteroposterior projection of the right ICA (D) and lateral projection of the left PcoA (E) show a retrograde flow into the left CCF.

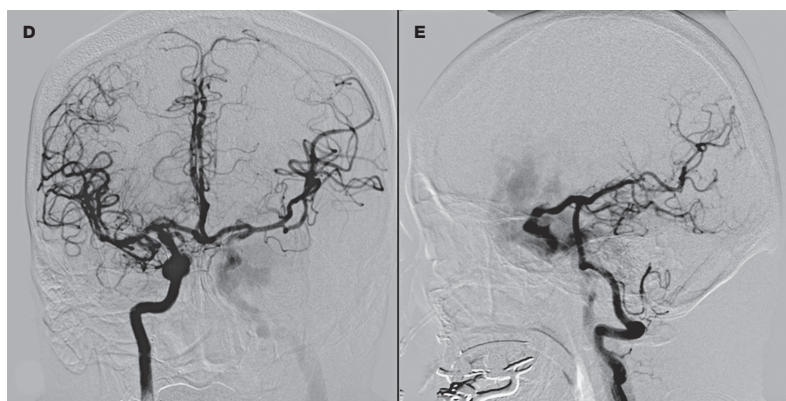


Figure 3 CT scan reveals an acute brainstem hemorrhage, causing mild obstructive hydrocephalus and minimal intraventricular hemorrhage.

(ICA) that had ruptured at its posterior aspect into the left cavernous sinus. The right ICA and left vertebral artery angiogram revealed a high velocity collateral flow through the circle of Willis. The left anterior and middle cerebral arteries were supplied by a well-developed contralateral circulation through the anterior com-

municating artery (AcoA) and the left posterior communicating artery (PcoA), from which a partial steal of the blood flow was also noted. Severe cortical venous reflux via the distended left sphenoparietal sinus, deep Sylvian veins and cortical veins draining into the sagittal sinus was visualized early in the arterial phase.

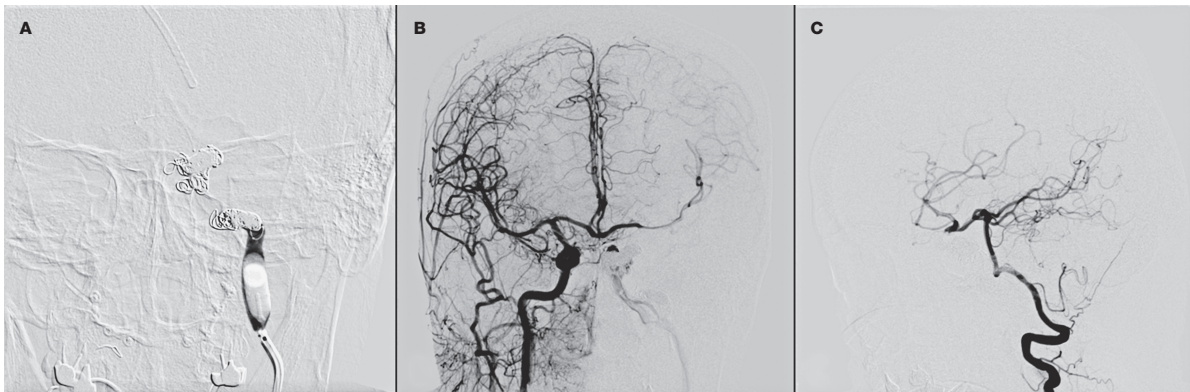


Figure 4 A-C) Left ICA angiograms during transarterial embolization (TAE). A) Guglielmi detachable coils and n-butylcyanoacrylate with temporary balloon assistance to sacrifice the left ICA. B, C) Post TAE angiograms show most parts of the CCF were obliterated and no obvious retrograde venous flow from the AcoA and PcoA.

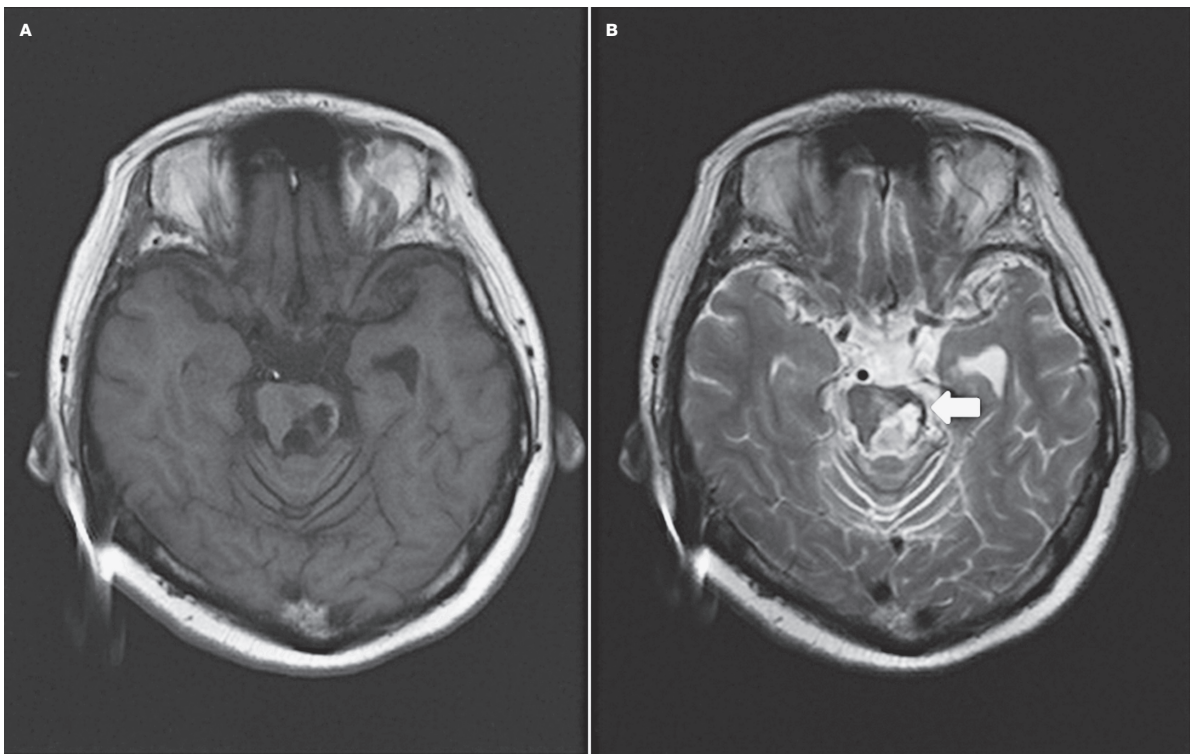


Figure 5 MRI, T1-weighted (A) and T2-weighted (B) images one year after treatment show a focal brain tissue loss with surrounding hemosiderin deposition at the left pons (B, arrow). There is no engorgement of veins surrounding the brainstem. Relative prominence of the left superior ophthalmic vein is noted but no definite residual or recurrent CCF could be seen.

The CCF also drained into the left inferior petrosal sinuses, causing a congestion of the brainstem and the anterior pontomesencephalic venous plexus. The left superior petrosal sinus was poorly visible. Significant venous congestion due to tortuous and dilated posterior fossa veins was also noted (Figure 2).

Initially, our treatment strategy of the CCF was stent-assisted coiling for fistula emboliza-

tion and left ICA preservation. But unfortunately, one day after the DSA, the patient became somnolent. An emergent brain CT scan showed a pontine hemorrhage, about 3×2.8×2.3 cm in size, causing minimal intraventricular hemorrhage (IVH) and mild obstructive hydrocephalus (Figure 3), which necessitated an emergent surgical intervention.

After right ventriculostomy for the IVH and

hydrocephalus, transarterial embolization (TAE) of the CCF and sacrifice of the left ICA just proximal and distal to the fistula site were attempted with continuous endovascular procedures. Initially, two microcatheters were advanced into the fistula via both the left proximal ICA and left vertebral artery-basilar artery-left PcoA-left distal ICA routes, followed by a temporary balloon assistance for the proximal left ICA flow control, and finally, embolization with Guglielmi detachable coils and n-butylcyanoacrylate. Completely obliteration of the CCF was accomplished right after the TAE (Figure 4).

Postoperatively, the patient's condition improved within one week, and she regained the same consciousness status she had on admission. The symptoms of headache and dizziness improved and there was no bruit over the left eye, but the left complete facial palsy and right hemiparesis persisted. The postoperative course was uneventful and the patient was discharged after one month in hospital. Three months after the embolization, DSA and MRI revealed no recurrence of the CCF, however, magnetic resonance imaging showed focal brain tissue loss with long T2 change on the left side pons and cerebral peduncle (Figure 5).

Discussion

Carotid-cavernous fistula (CCF) is an abnormal communication between the carotid artery and cavernous sinus. Etiological classification separates CCFs into traumatic CCF and spontaneous CCF. Traumatic CCF is the most common type, accounting for up to 75% of all CCFs. It was first described by Travers et al. in 1809. Barrow et al. defined four types of CCFs in 1985. Type A CCF is characterized by direct shunting of blood flow from the ICA into the cavernous sinus. Types B, C and D CCFs are all indirect low flow lesions that arise from the meningeal branches of either the external carotid artery or ICA.

The characteristic clinical features seen in patients with CCF depend on the direction of the drainage pattern. The classical symptoms include Dandy's triad of pulsatile exophthalmos, bruit and chemosis with anterior drainage via the ophthalmic vein. Uncommonly, with posterior drainage via the superior or inferior petrosal sinus to the jugular vein, elevated pressure in the posterior venous drainage may pro-

duce atypical symptoms including cranial palsy, hemiparesis and intracranial bleeding. In extremely rare cases, posterior venous drainage can result in brainstem congestion or hemorrhage via venous hypertension in the pre-pontine cistern or interpeduncular cistern.

Brainstem dysfunction with venous congestion or hemorrhage is a rare complication with CCF; only eight cases of direct CCF and 13 cases of indirect CCF have been reported as listed in Table 1¹⁻¹⁷. In most cases, brainstem dysfunction was caused by venous congestion, whereas only four cases, including ours, were found to have brainstem hemorrhage. The venous congestion by CCF usually involves the pontine parenchyma, midbrain, medulla oblongata or cerebellum. Venous congestion of the cervical spinal cord was found in two cases with direct/indirect CCF presenting with cervical myelopathy^{6,14}.

The pathomechanisms associated with venous congestion of the brainstem in CCF are still variable. Some authors supported the hypothesis of patterns of venous drainage being an important factor in the development of brainstem congestion. Murata et al. proposed that occlusion of the superior petrosal sinus by CCF causing engorgement of the veins in the surrounding brainstem could explain the development of brainstem congestion³. Ito et al.¹⁶ reported some mechanisms that may be the primary triggers for severe brainstem venous congestion: 1) a large flow volume shunted via the CCF, 2) occlusion of the superior or inferior petrosal sinus, 3) thrombosis of the dural sinuses or veins of the posterior fossa, and 4) variations in venous drainage. Table 1 shows that engorged veins were noted surrounding the brainstem in nearly every case of brainstem dysfunction due to a direct CCF. These characteristics were not seen in cases of indirect CCF. The presence of leptomenigeal venous drainage and of large variceal or venous pouch dilation in particular predicts an aggressive clinical behavior, such as intracranial hemorrhage, cerebral infarction and brain edema^{18,19}. Turner et al. surmised that a high flow CCF increased an abnormal back-pressure in venous drainage leading to a subsequent intracranial hemorrhage¹.

Some emergencies are immediately life-threatening: the presence of major external hemorrhage such as epistaxis, cortical venous drainage with symptoms of increased intracranial pressure, and angiographic demonstration of a venous pouch, have been described as indi-

Table 1 Brainstem dysfunction caused by carotid-cavernous fistula.

Case No.	Author	Year	Age/ Sex	Symptoms	Barrow's classification of CCFs	Drainage	Location	Hemorrhage	Congestion	Angiography: Engorged veins in surrounding brainstem	Treatment	Outcome
1	Turner et al	1983	81/F	OPT, Hemiparesis, Comatose	A	SOV, Sphenoparietal sinus	Midbrain	+		+	NA	Death
2	Ten et al	1991	36/M	Unconsciousness, Hemiparesis	A	Sphenoparietal sinus	Midbrain, Thalamus		+	+	TAE	Free
3	Ten et al	1991	26/M	Numbness of right side of body and face	A	SOV	Midbrain		+	+	TAE	Free
4	Murata et al	2003	49/F	OPT, Right hemiparesis	A	IPS	Pons		+	+	TAE	Free
5	Bussiere et al	2009	39/M	OPT, Left facial numbness, Dysarthria, Gait ataxia, Drowsy	A	SOV, IOV, PMC V.	Pons		+	+	TAE	Free
6	Aldea et al	2011	55/F	Left limbs weakness, Dysarthria, Confusion, Locked in state	A	SOV, SPS, IPS	Pons, Cerebellum	+		+	TAE	Free
7	Herrera et al	2011	27/M	Tetraparesis, Respiratory insufficiency	A	PMC V.	Medulla oblongata		+	+	TAE	Improved
8	Chan et al	2013	27/F	OPT, Left upper limb weakness, Paraplegia	A	SOV	Cervical spinal cord	+		NM	TAE	Neurological deficit
9	Uchino et al	1995	68/F	OPT, Urinary dysfunction, Left sided cerebellar ataxia	D	Basal V, V. of Labbe	Pons		+	-	TAE	Neurological deficit
10	Uchino et al	1995	74/M	OPT, Ataxic gait	D	Cortical V. of p. fossa	Pons, Cerebellum		+	-	TAE, Radiotherapy	Free
11	Takahashi et al	1999	49/M	OPT	C	SOV, IOV, SPS	Pons, Cerebellum		+	-	TVE	Improved
12	Takahashi et al	1999	62/F	OPT	D	SOV, IOV	Pons		+	-	TVE	Improved
13	Shintani et al	2000	65/F	OPT, Right hemiplegia	B	IPS	Pons, Medulla oblongata		+	-	NA	Death
14	Michael et al	2001	76/F	Dysarthria, Left hemiplegia	D	Petrosal V.	Midbrain		+	-	TAE	Free
15	Kai et al	2004	56/F	OPT, Left hemiparesis	D	Petrosal V.	Midbrain		+	-	Open surgery	Neurological deficit
16	Kai et al	2004	70/F	OPT, Gait instability, Ataxia	D	PMC V.	Midbrain		+	-	Open surgery	Free
17	Iwasaki et al	2005	71/F	OPT	C	SPS, PMC V.	Pons		+	-	Radiosurgery	Free
18	Ko et al	2008	54/M	OPT, Progressive quadripareisis	D	PMC V.	Medulla oblongata, Cervical spinal cord		+	-	TAE, Radiosurgery	Neurological deficit
19	Gierthmuehlen et al	2010	30/M	OPT, Somnolent, Anisocoria, Dysarthria	D	NM	Midbrain	direct compressed		-	TAE	Free
20	Ito et al	2011	76/F	OPT, Dysarthria, Gait instability	B	SOV	Pons		+	-	Endovascular treatment	Free
21	Miyagishima et al	2012	80/F	OPT, Ataxia, Dysarthria, Left hemiparesis	D	SOV, Petrosal V.	Pons		+	-	TAE, Radiation	Free
22	Present case	2013	34/F	OPT, Right hemiplegia, Left facial palsy, Comatose	A	SOV, Sphenoparietal sinus, IPS	Pons	+		+	TAE	Neurological deficit

NM: Not mentioned; NA: Not applicable; OPT: Ophthalmopathy; SOV: Superior orbital vein; IOV: Inferior orbital vein; PMC: Pontomesencephalic; V: Vein; SPS: Superior petrosal sinus; IPS: inferior petrosal sinus; TAE: Transarterial embolization; TVE: Transvenous embolization

cators of a high risk of bleeding that needs urgent intervention²⁰. In general, multitherapeutic endovascular approaches to CCFs include endovascular repair with detachable balloon occlusion, coil and material embolization, covered stent graft placement and a combination of these techniques²¹. Occasionally, a large lesion has less chance of ipsilateral carotid preservation; arterial sacrifice with parent artery occlusion is needed in life-threatening cases. Malan et al.²² classified post-traumatic CCF into small, medium and large fistulae based on angioarchitecture, which is helpful in selecting varying endovascular tools.

We found the venous pouch disruption in our patient was large and contained extensive thrombus. A large intracranial venous pouch consisting of mural thrombosis is extremely rare. Thrombus could only be found in the artery aneurysm sac. Some hypotheses claim that the aneurysm wall might be exposed to varying thrombus-inducing autolytic activities leading to aneurysm wall degradation with rupture. Fontaine et al.²³ reported that the mural thrombus acts as a site of protease cascade and increased chemical degradation and weakening of the aneurysm wall, while Kulcsár et al.²⁴ found that large aneurysms with rapidly accumulated thrombosis may be associated with an increased risk of delayed rupture. Unfortunately,

ly, there is no actual evidence to show a correlation between venous pouch thrombosis and artery aneurysm thrombosis. Our patient's clinical course was marked by a rapid progressive brainstem hemorrhage due to posterior fossa venous congestion. Not only can a direct CCF with high pressure and large flow volume induce hypertension in the veins surrounding the brainstem, but a large venous pouch with thrombosis can also increase the risk of hemorrhage. Further investigation may be needed to support this hypothesis.

Although the overall mobility and mortality of CCF are quite low, brainstem congestion or hemorrhage, without prompt treatment, is a life-threatening disorder. Two patients who died of brainstem compression and central failure without endovascular procedure treatment have been reported^{1,16}. Irreversible brain edema, congestive venous ischemia and hemorrhage caused by rupture of the congested veins, are the prognostic indicators in patients with brainstem dysfunction caused by CCF.

In conclusion, brainstem hemorrhage is a rare but potentially serious complication of CCF. It may indicate a poor prognosis with an irreversible neurologic deficit or life-threatening condition. Early diagnosis and prompt treatment are very important in cases of direct CCF with brainstem venous congestion.

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Cho-Shun Li, MD
Department of Medical Education (Neurosurgery)
Chung Shan Medical University Hospital, Taiwan
110, Sec.1, Chien-Kuo N. Road
Taichung City, Taiwan (40201)
Tel.: +886-4-24621372
Fax: +886-4-24621371
E-mail: fhchantw@gmail.com (Fook-How Chan)
hkli@ms28.hinet.net (Cho-Shun Li)