Spontaneous intracerebral hematoma in carotid-cavernous fistula

Report of three cases

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Spontaneous intracerebral hematoma associated with carotid-cavernous fistula is rare. Three new cases are presented. In each, the hemorrhage originated in the vicinity of localized intracranial venous engorgement, as demonstrated by cerebral angiography. Rupture of one or several of the distended venous channels from increased back-flow is postulated as the etiology of the intraparenchymal hematomas.

Case Reports

Case 1

This 68-year-old man fell down a flight of stairs on November 7, 1981, suffering temporary loss of consciousness and multiple facial abrasions. Four days later, he noticed the gradual onset of severe orbital pain, and protrusion and congestion of the right eye.

Examination. On November 17, the patient's visual acuity was 20/60 in the right eye and 20/20 in the left, with intraocular pressures of 30 and 15 mm Hg, respectively. The right eye was proptotic and pulsatile, and a loud bruit was auscultated over the right orbit. There was an oculomotor and trochlear nerve paresis with an abducens nerve paralysis on the right, associated with a depressed corneal reflex. Computerized tomographic (CT) examination of the head demonstrated proptosis of the right globe, an area of increased density in the posterior portion of the right orbit within muscle, and a small right-sided low-density subdural hematoma with minimal mass effect. Right carotid angiography demonstrated a high-flow right-sided CCF, with engorgement and rapid filling of the right superior ophthalmic vein and its tributaries. Dural and subfrontal pial veins were distended, and the cortical veins draining into the sagittal sinus were visualized early in the arterial phase (Fig. 1).

The patient was admitted to University Hospitals on November 20, and the pertinent physical findings were intense chemosis, periorbital edema, and 7-mm proptosis of the pulsating right eye. A loud synchronous bruit was heard over the right orbit, and there was total ophthalmoplegia with a decreased corneal reflex. Visual acuity in the right eye was limited to finger counting only. Three days later, he suddenly lost consciousness and exhibited a left hemiparesis. A CT scan demonstrated a large frontal intracerebral hematoma on the right side (Fig. 2). Right carotid angiography demonstrated the findings as noted previously (Fig. 3). In addition, there was posterior displacement of the Syl-
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FIG. 1. Case 1. Prehemorrhage right carotid subtraction angiograms, anteroposterior (left) and lateral (right) views. There is a high-flow right carotid-cavernous fistula (large open arrow) with engorgement and rapid filling of the right superior ophthalmic vein (small open arrow) and its tributaries. Engorgement of the subfrontal pial and dural veins is also seen (large closed arrow). The right frontal cortical veins are visualized early in the arterial phase (small closed arrows), with early filling of the superior sagittal sinus.

vian triangle and stretching of the frontal opercular branches of the middle cerebral artery. There was significant venous drainage through the clival-basal venous plexus.

Operation. The intraparenchymal and intraventricular hematomas were removed via a right frontotemporal craniotomy. Diffuse bleeding was identified from intraparenchymal distended subfrontal veins, which were thought to be the origin of the intracerebral hematoma. The right CCF was occluded by retrograde packing of the common ophthalmic vein, and there was immediate resolution of the right orbital bruit. A right ventriculostomy was performed for cerebrospinal fluid drainage.

Postoperative Course. Immediately after operation, the proptosis and chemosis receded and the ventriculostomy tube was removed after 5 days. The patient’s mental status and left hemiparesis gradually improved, and postoperative carotid angiography on January 8, 1982, showed the fistula to be totally obliterated; the internal carotid artery was patent. Six months after discharge, the patient’s mental status was dull and a mild left hemiparesis was evident. Extraocular movements on the right were normal and there was complete resolution of the chemosis and proptosis; however, visual acuity was unchanged, with finger counting only.

Case 2

This 54-year-old man was involved in an automobile accident on March 24, 1979. During the subsequent 10 weeks, he developed symptoms of bilateral sixth nerve paresis, greater on the left than on the right, a left orbital bruit, chemosis, and proptosis of the left eye. This was followed by a sudden onset of expressive aphasia and right arm paresis on June 6, 1979, which precipitated admission to the hospital in Lincoln, Nebraska. A CT scan revealed a left frontotemporal intracerebral hematoma (Fig. 4).

Six days after his ictus, bilateral carotid and left vertebral angiograms demonstrated a high-flow CCF on the left, with early filling and distention of the left
FIG. 3. Case 1. Posthemorrhage right carotid angiograms, anteroposterior (left) and lateral (right) views. The high-flow right carotid-cavernous fistula (large open arrow), right superior ophthalmic vein (small open arrow), and subfrontal pial and dural veins (closed arrows) are again visualized as in Fig. 1. In addition to the previous findings, there is now evidence of mass effect in the frontal lobe as shown by posterior displacement of the Sylvian triangle and stretching of the frontal opercular branches of the right middle cerebral artery.

FIG. 4. Case 2. Computerized tomography demonstrating a left posterior frontal intracerebral hematoma without mass effect.

superior ophthalmic vein and cortical veins which drain into the superior sagittal sinus. There was early visualization of multiple tortuous veins in the brain parenchyma persisting into the late arterial phase. These veins were located predominantly in the left frontal and left anterior parietal regions. A left frontal mass effect was also present (Fig. 5). The next day, the patient suffered a second ictus with an increase in his right hemiparesis and aphasia. Repeat CT scanning showed enlargement of the previous intracerebral hematoma (Fig. 6).

Operation. The CCF was occluded with a balloon using a No. 8 Foley Silastic catheter, and the left internal carotid artery was ligated in the neck. Intraoperative cerebral angiography revealed retrograde flow in the internal carotid artery distal to the fistula. Immediately after the balloon was inflated with Conray-60, the left orbital bruit disappeared. No effort was made to remove the intracranial hematoma.

Postoperative Course. The fistula remains obliterated 3 years after surgery. The patient has a mild aphasia with poor memory, but the abducens nerve palsies, chemosis, proptosis, and the right hemiparesis have resolved.

Case 3

This 81-year-old woman developed the acute onset of spontaneous severe bilateral retro-orbital pain on February 20, 1980. This was followed several hours later by bilateral periorbital edema, chemosis, and decrease in vision.

Examination. Physical examination on February 28 showed bilateral proptosis, a trace of light perception in the left eye, no light perception in the right, bilateral ophthalmoparesis, and a bruit over the right orbit. Bilateral carotid angiography revealed a high-flow right CCF. The right carotid angiogram demonstrated rapid filling of the right superior ophthalmic vein and cross-filling to the left cavernous sinus. Left cortical veins draining into the superior sagittal sinus and the left jugular vein filled early in the arterial phase. There was engorgement of the anterior pontomesencephalic vein and the veins in the rostral prepontine and interpeduncular cisterns (Fig. 7).

On March 21, 1981, the patient suddenly developed
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Fig. 5. Case 2. Left carotid angiograms, anteroposterior (left) and lateral (right) views, demonstrating a high-flow left-sided carotid-cavernous fistula (large open arrow) with early filling and distention of the left superior ophthalmic vein (small open arrow). Early filling of the left frontal cortical veins (small closed arrows) draining into the superior sagittal sinus is seen. There is extensive early visualization of many small tortuous venous structures within the brain parenchyma (large closed arrows) that remain filled until the late arterial phase and are predominantly localized to the frontal and anterior parietal regions. Mild evidence of left frontal mass effect is present.

A right hemiparesis and became comatose. A CT scan revealed a left midbrain hemorrhage (Fig. 8). She continued to deteriorate and died 1 month later.

Discussion

Since the first clinical description of “pulsating exophthalmos” by Travers in 1809, CCF has been a well studied and frequently reported entity. Its incidence is estimated to be one case among 20,000 hospital admissions, and it is the most common site of arteriovenous aneurysm because of the unique anatomy of an artery passing through a venous channel. The older literature has referred to CCF by a variety of terminologies including “erectile tumor” of the orbit, “aneurysm-by-anastomosis,” “diffuse aneurysm of the ophthalmic artery,” “arteriovenous anastomosis,” “pulsating exophthalmos,” and “carotid cavernous aneurysm.”

The cardinal symptoms of a CCF are exophthalmos, ocular pulsation, and orbital bruit. Visual loss has been estimated to occur in 25% of cases, and visual impairment in an additional 20%. Other symptoms and signs have been described, including ophthalmoplegia, chemosis, ocular pain, hypalgesia in the first division of the trigeminal nerve, and absence of the corneal reflex. In general, CCF has been considered as not life-threatening, and the goal of therapy has been “preservation or improvement of vision, return of the orbit and its contents to normal, and elimination of the bruit.”

Spontaneous hemorrhage from CCF is rare. In 1908, deSchweinitz and Holloway reported the first case of
FlG. 7. Case 3. Right carotid angiogram, anteroposterior (left) and lateral (right) views, demonstrating a high-flow right carotid-cavernous fistula (large open arrows) with early filling and engorgement of the right superior ophthalmic vein (small open arrows). Rapid cross-filling of the left cavernous sinus (other large open arrow) is evident. Left: Filling of the left cortical veins (small closed arrow) and left jugular vein (large closed arrow) can be seen prior to any filling of the venous structures on the right. Right: Lateral view demonstrates engorgement of the anterior pontomesencephalic vein and the veins in the vicinity of the rostral preptontine and interpeduncular cisterns (closed arrow).

FIG. 8. Case 3. Computerized tomography scan displaying an area of hemorrhage near the left midbrain and left hypothalamic-thalamic regions.

Intracerebral hematoma in a patient with traumatic CCF. At autopsy, the right cavernous sinus had ruptured at the extreme posterior area immediately adjacent to the apex of the petrous portion of the temporal bone. In 1930, Sattler added two additional cases of intracerebral hematoma secondary to rupture of the cavernous sinus in both a spontaneous and traumatic fistula. He reviewed 322 previous cases of CCF, and reported five fatal cases of epistaxis and three intracerebral hematomas, citing an incidence of 1.5% and 0.9%, respectively. Based on his manuscript, a 3% incidence of spontaneous hemorrhage associated with CCF has been quoted. Subsequently, Ambler, et al., described a single case of CCF exhibiting multifocal petechial and elliptical hemorrhages in addition to a circumscribed intraparenchymal hematoma. The hemorrhagic lesions were attributed to either local arteriovenous shunting or to remote arterial steal combined with venous hypertension secondary to reflux into the venous channels draining the cavernous sinus.

In addition to the five cases of fatal epistaxis following rupture of a CCF reported by Sattler, other cases of epistaxis have been documented or suggested in association with this entity, although such bleeding is more commonly observed following rupture of a traumatic intercavernous carotid artery aneurysm. A single case of conjunctival hemorrhage associated with CCF has also been described, as well as two cases of subarachnoid hemorrhage. Bartlow and Penn reported a patient with a cerebellopontine angle mass lesion secondary to aneurysmal dilatation and thrombosis of the superior petrosal vein from a CCF.

In each of our three cases, spontaneous intracerebral hemorrhage occurred at those sites in the brain where venous engorgement was evident on cerebral angiography. Case 1 demonstrated a high-flow CCF with venous engorgement of the right subfrontal pial and dural veins, and the frontal cortical veins draining into the superior sagittal sinus on the involved side were visualized early in the arterial phase. At operation to evacuate the spontaneous intracerebral hematoma, bleeding appeared to originate from ruptured subfrontal pial veins.

In Case 2, there was extensive early visualization of...
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multiple small tortuous veins within the brain parenchyma, which remained filled through the late arterial phase. These were localized within the left frontal and anterior parietal regions. The left cortical veins and superior sagittal sinus were visualized early. There were no angiographic vascular abnormalities evident in the right hemisphere. The spontaneous hemorrhages occurred on two different occasions in this case and were within the left posterior frontal region at the sites of angiographically demonstrated venous hypertension.

Case 3 showed a high-flow right CCF with rapid cross-filling to the left cavernous sinus. There was visualization of the left cortical veins and left jugular vein before the right. There was engorgement of the anterior pontomesencephalic vein and the veins in the vicinity of the rostral preponine and interpeduncular cisterns. The area of hemorrhage in this case was localized to the left midbrain area.

Although multiple routes of venous drainage from the cavernous sinus to the internal jugular vein have previously been described, spontaneous intracerebral hemorrhage has not previously been linked to angiographically demonstrated focal areas of venous hypertension. We conclude from these three cases that the increased abnormal back-pressure in veins draining the high-pressure CCF was the cause of spontaneous rupture of one or several of those abnormally distended venous channels, leading to spontaneous intracerebral hematoma.

References

24. Martin JD Jr, Mabon RF: Pulsating exophthalmos. Re- view of all reported cases. JAMA 121:330-335, 1943

J. Neurosurg. / Volume 59 / October, 1983
41. Travers B: A case of aneurysm by anastomosis in the orbit cured by ligation of the common carotid artery. Trans Med Chir Soc 2:1–16, 1809

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