Bilateral carotid-cavernous fistulae of mixed types with unusual radiological and neuropathological findings

Case report

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This report describes a case of bilateral post-traumatic carotid-cavernous fistulae (CCF), of both typical and atypical types, with delayed clinical deterioration. Unusual neuropathological lesions, distinctive from those due to direct cerebral trauma, are related to combined arterial ischemia and venous hypertension. Atypical CCF is not necessarily a benign disorder. Radiological monitoring is essential to detect spontaneous progressive intracranial shunting, to predict areas that are at risk from venous hypertension, and to identify remote sites of circulatory vulnerability.

KEY WORDS • carotid-cavernous fistula • venous hypertension • radiology • neuropathology

CAROTID-CAVERNOUS sinus fistula (CCF) is most commonly unilateral and is usually caused by head trauma.6 The dramatic ocular-orbital symptoms are principally due to orbital venous hypertension. Ocular necrosis is a serious potential complication.7,8 Surgical treatment designed to reduce carotid arterial flow carries the additional risk of cerebral ischemia, which is rarely symptomatic in untreated patients.10

In some instances of CCF, venous filling of intracranial dural sinuses and intracerebral veins is prominent. This atypical pattern, more often seen in nontraumatic fistulae, is considered a low-pressure alternative venous drainage of a benign consequence.5,11 A single case of atypical CCF with subarachnoid hemorrhage attributed to cortical venous hypertension has been recorded.6 Cerebral parenchymal lesions resulting from intracranial venous hypertension have not been described.

The patient reported here had bilateral post-traumatic carotid-cavernous fistulae of mixed types; she suffered spontaneous neurological deterioration and died 4 months following the head injury. Radiological demonstration of progressive intracranial arterial insufficiency and high-pressure venous reflux is correlated with unusual cerebral parenchymal and vascular lesions.

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This 64-year-old woman was admitted December 6, 1974, following an automobile accident that resulted in a severe head injury. She complained of blurred vision and ocular motility disturbances, as well as bilateral proptosis. Radiological studies revealed bilateral carotid-cavernous fistulae, with prominent venous filling of the intracranial venous system. Despite initial medical treatment, her neurological status deteriorated progressively, and she died 4 months later.

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accident. She was unresponsive, cyanotic, and hypotensive with a flail chest. Random right eye movements and sporadic limb movements were noted. The left eye was protruded and immobile with a dilated and fixed pupil. Improvement of color and vital signs followed the treatment of her pneumothorax. Further examination showed a right postauricular laceration with blood and spinal fluid exuding from the right ear. Skull and cervical spine x-ray films did not demonstrate fractures. An echoencephalogram showed no midline deviation. Further x-ray films revealed fractures of a clavicle, ribs, and ankles and feet. Following treatment with Osmitrol (mannitol) and Decadron (dexamethasone) she was more alert and attended to command, but was aphasic. Purposeful movements were preserved, but were more facile on the left. Bilateral hyperreflexia was noted.

Ocular evaluation showed a normal right eye with a 2-mm pupil and intact ocular mobility. The left eye showed ptosis, chemosis, ophthalmoplegia, and a fixed, dilated pupil. The left facial veins were distended. Because of bilateral orbital bruits and progressive left exophthalmos, bilateral carotid angiography was performed on December 28, and demonstrated bilateral carotid-cavernous fistulae with considerable variation in venous reflux. Direct extracranial flow from the cavernous sinuses into the ophthalmic veins was shown bilaterally (Fig. 1) and into the pterygoid plexus on the right (Fig. 1 upper). Intracranially, the inferior petrosal sinuses provided drainage bilaterally, and there was crossover to the opposite cavernous sinus during the left injection (Fig. 1 lower left). On the right, there was prominent reflux into the sphenoparietal sinus and the middle cerebral vein and their tributaries in the cerebral hemisphere (Fig. 1 upper). This was associated with early opacification of the cortical veins draining into the superior sagittal sinus. Reflux from the right cavernous sinus also occurred into veins at the base of the brain which communicated via a network of small vessels with the vein of Rosenthal, vein of Galen, and straight sinus (Fig. 1 upper right). A small vein and a few of its tributaries were also shown in the sphenoparietal area on the left (Fig. 1 lower right).

The ipsilateral, anterior, and middle cerebral arterial flow was preserved bilaterally, despite the proximal fistulae. The right vertebral injection opacified both posterior cerebral arteries, but also showed prominent flow through both posterior communicating arteries to the left and right anterior and middle cerebral arteries (Fig. 2).

Drainage of contrast material away from the left carotid-cavernous fistula was classical. On the right, drainage was demonstrated into the pterygoid plexus as well as into ophthalmic veins. In addition, there was unusual and prominent high-pressure reflux into the middle cerebral vein and veins at the base of the brain which are normally efferent to the cavernous sinus. Both carotid arteries continued to supply their respective anterior and middle cerebral branches. Nevertheless, there was such prominent forward flow through both posterior communicating vessels during the

Radiological Findings

First Examination. Bilateral carotid and right vertebral injections on December 28, 1974, demonstrated bilateral carotid-cavernous fistulae with considerable variation in venous reflux. Direct extracranial flow from the cavernous sinuses into the ophthalmic veins was shown bilaterally (Fig. 1) and into the pterygoid plexus on the right (Fig. 1 upper). Intracranially, the inferior petrosal sinuses provided drainage bilaterally, and there was crossover to the opposite cavernous sinus during the left injection (Fig. 1 lower left). On the right, there was prominent reflux into the sphenoparietal sinus and the middle cerebral vein and their tributaries in the cerebral hemisphere (Fig. 1 upper). This was associated with early opacification of the cortical veins draining into the superior sagittal sinus. Reflux from the right cavernous sinus also occurred into veins at the base of the brain which communicated via a network of small vessels with the vein of Rosenthal, vein of Galen, and straight sinus (Fig. 1 upper right). A small vein and a few of its tributaries were also shown in the sphenoparietal area on the left (Fig. 1 lower right).

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Fig. 1. Upper: Anteroposterior (left) and lateral (right) views of right carotid arteriogram. Lower: Anteroposterior (left) and lateral (right) views of left carotid arteriogram. Long solid arrows: ophthalmic veins and pterygoid plexus; small arrows: sphenoparietal sinus and middle cerebral vein and tributaries; curved solid arrows: inferior petrosal sinus; open arrowheads: basal vein and tributaries, vein of Galen, and straight sinus; open straight arrows: cortical veins; open curved arrows: jugular vein; solid arrowheads: cavernous sinus crossover. There is good opacification of anterior and middle cerebral arteries bilaterally, in spite of the proximal fistulae. Note that, in these arterial phase films, much of the opacified intracranial vasculature on the right is venous.

Vertebral study that it probably represented an arterial "steal" phenomenon from the vertebrobasilar system. 12

Second Examination. Bilateral carotid arteriography on January 30, 1975, showed that in the month interval little change had occurred on the left side. Crossover to the right cavernous sinus was no longer present. The anterior and middle cerebral arteries were again opacified.

There was, however, marked change on the right (Fig. 3). Extracranial flow into the ophthalmic veins was decreased, and the pterygoid plexus was not opacified. There was prominent increase of reflux into the sphenoparietal sinus, middle cerebral vein, and their tributaries in the right hemisphere, and into the veins at the base of the brain, accompanied by earlier and denser opacification of cerebral cortical veins and of the veins of
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die cerebral vein and its tributaries, and into veins at the base of the brain. In this final study, visualization of these veins was enhanced by complete absence of opacification in the anterior and middle cerebral arteries. All of the internal carotid flow had become diverted into the cavernous sinus, and thence into cerebral veins and dural sinuses. A vein that bled profusely at the time of surgery for suspected subdural hematoma and was clipped is shown to have been the middle cerebral vein. The unusual bleeding is attributable to high-pressure reflux from the carotid artery.

**Autopsy Findings**

Necropsy was restricted to the head. Recent right-sided burr holes were associated with local subdural and subarachnoid hemorrhage with adjacent cortical necrosis. The brain weighed 1145 gm. The right parasylvian gyri, preserved in outline, were swollen and soft. There was subfalcral and right tentorial herniation with right inferior temporal hemorrhagic infarction due to posterior cerebral artery compression. Multiple old contusions were seen on the left orbital and anterior temporal surfaces. The cavernous, sagittal, transverse, and sigmoid sinuses were patent. No skull fractures were observed.

**Third Examination.** The right carotid angiogram on March 7, 1975, demonstrated progression of the changes seen on the second examination (Fig. 4). There was no longer any reflux into the right ophthalmic veins. On the other hand, marked retrograde flow continued into the sphenoparietal sinus, the middle cerebral vein and its tributaries, and into veins at the base of the brain. In this final study, visualization of these veins was enhanced by complete absence of opacification in the anterior and middle cerebral arteries. All of the internal carotid flow had become diverted into the cavernous sinus, and thence into cerebral veins and dural sinuses. A vein that bled profusely at the time of surgery for suspected subdural hematoma and was clipped is shown to have been the middle cerebral vein. The unusual bleeding is attributable to high-pressure reflux from the carotid artery.

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Cross sections of the brain showed multiple left (contrecoup) orbital and temporal contusions. A restricted area of cortical swelling and granularity involved orbital and temporal borders of the proximal left Sylvian fissure (Fig. 5).

In the right hemisphere, a large wedge-shaped area of "tumorous" swelling involved the third frontal gyrus, all of the insular cortex, and the entire anterior temporal lobe extending medially to the lateral striatal margin (Fig. 5 left). The gyral contour was maintained, but cortical demarcation was obscured. The white matter was swollen and boggy with multiple petechial and elliptical hemorrhages. There was no cavitary degeneration. Sylvian vessels were thickened (Fig. 5 left). Posteriorly, a large concentrically lobulated hematoma was located in the softened suprasylvian white matter.

Midbrain sections contained circumscribed tegmental and paramedian necrosis due to herniation. The rostral basal pons showed right-sided swelling, softening, and granularity with petechial hemorrhages (Fig. 5 right) involving the entire basis pontis at lower levels. The medulla was normal.

Microscopic Examination

In the cavernous sinus, arterial intimal thickening was noted without demonstration...
of direct arteriovenous communications. The venous walls were thickened, and fibrous proliferation extended into the cranial nerves and Gasserian ganglia with focal demyelination.

In the brain, bilateral contusions, circular and laminar cortical infarcts seen in frontotemporal and orbital areas mainly on the left, were attributed to the original head injury. The “tumorous” softenings in both hemispheres and basal pons were accompanied by marked vascular alterations not observed in the unaffected areas. Surface arteries were patent with minimal subintimal proliferation. Pial veins were profoundly hypertrophied with mural fibrous hyperplasia (Fig. 6 left). In the hemispheric lesions there was marked cortical cell loss and replacement gliosis. Edematous vacuolization, microvascular thickening, and petechial hemorrhages were most frequent in white matter (Fig. 6 right).

In the vicinity of the right subcortical hematoma there were foci of pigmented macrophages and thick vessels with both old and recent thrombi. The basal pons showed myelinolysis, fibrous gliosis, edema, and petechial extravasation with similar thickening of surface and penetrating veins (Fig. 7).

Discussion

In typical CCF, despite significant shunting of internal carotid flow, intracranial complications are relatively rare in untreated cases, probably because the disorder is unilateral and the cerebral collateral circulation is competent. The major organ at risk is the eye. Ocular necrosis differs from pure ischemic damage and is attributed to both
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reduced arterial perfusion and the congestion, edema, and extravasation accompanying venous hypertension. The patient described above showed unusual multifocal parenchymal and vascular lesions of the brain of similar dual circulatory origin. The multifocal cerebral and pontine lesions differ from those of either arterial or venous occlusive disease. Venous hypertrophy limited to involved areas resembles the mural alterations described in veins subjected to high luminal pressure.

Radiological findings support this pathogenesis and provide the basis for the multiple locations and varying extent of the lesions. On the right there was initial carotid arterial shunting into both extracranial and intracranial venous channels. Subsequent studies demonstrated progressive high-pressure reflux into intracerebral veins and venous sinuses as well as diminished intracerebral arterial opacification. The major lesion in the parasympathetic region was attributed to combined arterial ischemia and venous hypertension. Failure to demonstrate parenchymal lesions in other portions of the right hemisphere was probably the result of effective arterial collateral flow from the posterior circulation. The radiological impression that opacification of superior cortical veins represented normal venous drainage is supported by the lack of "arterialization" of these channels.

On the left, typical CCF was associated with predominantly extracranial orbital venous reflux. The limited cerebral lesion in the proximal Sylvian area is explained by the less severe arterial insufficiency and minimal reflux into middle cerebral veins noted in this area.

The pontine lesion of similar character was more difficult to explain since direct arteriovenous shunting did not affect this area. Initial vertebral angiography showed supratentorial diversion of the posterior circulation. This arterial steal likely progressed with the increased right carotid-cavernous shunting resulting in chronic pontine ischemia. The transverse pontine veins normally drain via the basal venous plexus into the cavernous sinus, proving an anatomical route for potential venous flow reversal.

Mural hypertrophy of pontine veins confirms the role of venous hypertension in the pathogenesis of the pontine lesion.

During the final admission, because of the history of head trauma, delayed clinical deterioration and prominent midline echo shift, the presumptive diagnosis of subdural hematoma was made; however, burl-hole exploration produced negative results. In reality, the terminal course is attributable to chronic venous hypertension, intracerebral venous hemorrhage, and internal cerebral herniations.

Although the present case is unusually complicated with bilateral fistulae of mixed types, it is particularly helpful in emphasizing the inherent dangers of the atypical form of CCF. In the absence of alarming ocular-orbital symptoms, progressive intracranial arterial insufficiency accompanied by simultaneous venous hypertension may cause neurological deterioration and death. The cerebral lesions, distinctive grossly and microscopically, may be multiple due to either the local arteriovenous shunting or to remote arterial steal combined with reflux into any of the venous channels connecting with the cavernous sinus. Sequential radiological studies in atypical CCF are important to detect unusual and progressive hemodynamic alterations, distinguish high-pressure venous reflux from low-pressure alternative drainage, and identify unsuspected remote areas of dual circulatory vulnerability.

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References


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