Detection of cryptic vascular malformations by computerized tomography

Report of two cases

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Two cases of cryptic vascular malformation that were not demonstrated by cerebral angiography were detected by computerized tomography. One of these patients had a cavernous angioma in the fourth ventricle with recurrent subarachnoid hemorrhages, and the other harbored a small arteriovenous malformation and intracerebral hematoma. The usefulness and limitations of computerized tomography in the identification of cryptic vascular malformations are discussed.

KEY WORDS - cryptic arteriovenous malformation - cavernous angioma - spontaneous intracerebral hematoma - computerized tomography

Cryptic vascular malformations are not a frequent cause of intracerebral or subarachnoid hemorrhage (SAH), and are rarely demonstrated by angiography. Computerized tomography (CT), however, holds great promise as an effective, noninvasive means of diagnosing this entity. Since April, 1977, two cases of cryptic angioma with intracerebral hematoma or SAH were detected by CT in our hospital. The vascular malformations were surgically removed and studied histologically. During the same period, six cases of spontaneous intracerebral hematomas and seven cases of SAH with unknown origin were encountered.

Case Reports

Case 1

This 29-year-old woman was admitted to our hospital on April 25, 1976, complaining of severe headache, sudden dizziness, nausea, and vomiting. Three months before this admission, the patient had suffered a similar attack of severe headache, nausea, and vomiting, and was admitted to another hospital, but details of the examination and treatment were not available.

Admissions. On admission, the patient was obtunded, and had a stiff neck; otherwise neurological findings were unremarkable. Lumbar puncture revealed markedly blood-stained cerebrospinal fluid (CSF) under a pressure of 260 mm H₂O. Selective four-vessel serial angiography was carried out by the transfemoral route, but no pathological causes for the SAH, such as aneurysm or angioma, were detected. In addition, selective external carotid angiography and spinal angiography by catheterization failed to reveal the source of the bleeding. Hypertension was not present during her hospital stay, nor in her past history. Laboratory examinations, including blood coagulation studies, were all normal. With conservative treatment, she gradually improved and was free of symptoms at the time of her discharge on June 12, with a diagnosis of SAH of unknown origin.

On January 3, 1977, the patient had a third episode of SAH, and was brought to the emergency service. Selective catheterization of the right and left carotid arteries and left vertebral artery again failed to reveal any vascular lesion. Technetium-99m brain scan and pneumoencephalography were carried out but demonstrated no abnormality. She was discharged again on March 25, 1977, without the causative lesion being identified.

On July 10, 1977, the patient had a fourth SAH and was readmitted. A plain CT scan revealed a small, poorly circumscribed area with increased density on the left wall of the fourth ventricle (Fig. 1 left).
area was enhanced by a rapid intravenous injection of 100 ml of 65% meglumine iothalamate. The fourth ventricle appeared to be deformed by a mass (Fig. 1 right), which showed an average of 35.4 Hounsfield units in the plain scan and 62.8 in the enhanced scan. Vertebral angiograms including subtraction films were rechecked retrospectively, but no lesion compatible with the CT findings was identified. Serial vertebral angiography with a magnification factor of × 2 was performed, but again demonstrated no vascular lesion in the fourth ventricle (Fig. 2). The clinical course with recurrent SAH's and the CT findings led to a tentative diagnosis of a small vascular malformation of cavernous or telangiectatic type. Operative intervention was recommended, but due to the patient's unavoidable circumstances, surgery was deferred until May 16, 1978, when suboccipital craniectomy was performed.

Operation. After an incision in the posterior part of the vermis, the fourth ventricle was explored under microsurgical technique. A small lobulated vascular mass, purplish-red in color, was found on the ventrolateral wall of the fourth ventricle. Orange discoloration abutting the mass suggested that it had bled previously. The mass, which was like a mulberry in appearance and measured 1.0 × 1.0 × 0.8 cm, was removed totally with very little bleeding. Postoperatively, dizziness and unsteady gait persisted over the next 2 weeks, but diminished gradually, and the patient was discharged on June 15 with no neurological deficits.

Histologically, the mass was composed of closely packed venous channels showing a typical honeycomb appearance and venous reservoirs with wide lumen (Fig. 3). Calcification or thrombosis was not demonstrated. The histological diagnosis was cavernous angioma.

Case 2

This 60-year-old man had an attack of severe headache, nausea, vomiting, and disturbance of consciousness on October 20, 1977. He was taken to another hospital, where lumbar puncture yielded...
blood-tinged fluid with elevated CSF pressure. Within a week, his consciousness gradually returned to normal, but he complained of a visual field defect. He was transferred to our hospital for further evaluation on November 10.

**Examination.** On admission, neurological examination disclosed right homonymous hemianopia and slight fluent aphasia, but the remainder of the examination was normal. Blood pressure was 140/90 mm Hg. There was no history of hypertension. All laboratory data including blood coagulation studies were within the normal range. Selective catheterization of the right and left internal carotid arteries and the left vertebral artery was performed. Angiograms revealed an intracerebral mass in the left precuneal region, but no aneurysm nor vascular malformation. A plain CT scan showed an intracerebral hematoma in the left parieto-occipital region (Fig. 4 left), and a CT scan enhanced with intravenous injection of 100 ml of 65% meglumine iothalamate showed a typical circular blush surrounding the hematoma. On the medial posterior part of the ring blush, a small spotty area with higher density was visualized (Fig. 4 right). The ring blush showed an average of 60 Hounsfield units and the spotty area, 78. Cryptic angioma was suspected. Serial angiography of the left internal carotid artery and vertebral artery was carried out, with a magnification factor ×2, but no vascular abnormality was demonstrated (Fig. 5).

**Operation.** Craniotomy was performed on November 29. After removal of intracerebral hematoma through a corticotomy in the left parieto-occipital junction, the hematoma cavity was washed repeatedly with saline, and the wall of the hematoma cavity was carefully explored. On the medial and inferior part of the wall, corresponding to the enhanced spotty area on the CT scan, a small, grossly lobulated, conglomerate of vessels was found. At least two feeding arteries and a draining vein were identified under the surgical microscope and were electrocoagulated. A vascular mass, approximately 1 cm in diameter, was removed without difficulty. The patient's postoperative course was uneventful. His speech disturbance and homonymous hemianopia improved, and he was discharged on December 22 in good condition.

Histologically, the mass was composed of a mixture of many arterial vessels of various sizes and
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venous vessels with thickened walls or arterialized veins. No evidence of thrombotic changes of the vessels was seen. The pathological diagnosis was that of a small arteriovenous malformation (AVM) (Fig. 6).

Discussion

The term "cryptic vascular malformation" was first used by Crawford and Russell in 1956, to denote a small angioma, an infrequent cause of spontaneous intracerebral hemorrhage, which is not detectable by routine angiography or other clinical examinations. Since then, this ill defined term, as well as several other, similarly arbitrary terms, such as "microangioma" or "small vascular malformation," became widely used. Advances in technique, equipment, and modality of cerebral angiography, including rapid serial angiography, magnification angiography, angiotomography, and film-subtraction technique has made possible detection of some "cryptic" vascular malformations. In the era of CT scanning, it is reasonable to expect many previously cryptic vascular malformations to become visible. The usefulness of CT in the diagnosis of AVM's of average size has been well documented. However, documentation of the use of CT in the diagnosis of cryptic angioma is rare.

Cavernous angioma is a common histological type of cryptic vascular malformation. Angiographic identification of this anomaly has been thought to be difficult, but recently several authors have described its angiographic characteristics. These are: 1) absence of any enlarged feeding artery; 2) possibly an avascular mass in the arterial phase; 3) capillary blush in the capillary phase; and 4) dilated veins in the early venous phase. These angiographic characteristics of cavernous angioma are related to its pathophysiological peculiarity. According to Diamond, et al., cavernous angiomas are shunted on the venous side of the circulation and contain a large reservoir of slowly flowing blood; absence of accumulation of contrast medium may be due to dilution of the medium in the reservoir. On the other hand, the draining veins, which receive the blood both from the normal tissue and the malformation would contain a higher concentration of the contrast medium and could conceivably be demonstrated.

In our Case 1, the cavernous angioma was visualized by CT scan as a enhanced mass with an average of 62.8 Hounsfield units, but on a careful check of the angiograms with the above angiographic characteristics in mind, we failed to identify a cavernous angioma.

A cryptic angioma accompanied by intracerebral hematoma presents some difficulties in interpretation of the CT scan. During the process of resolution and absorption of intracerebral hemorrhage, various patterns of increased attenuation areas appear with or without enhancement, which might be described as a blush, speckled, or moth-eaten. Some of these patterns are similar to or very often indistinguishable from those seen with a cryptic angioma which was responsible for the hemorrhage. Furthermore, the pathophysiological background for appearance and decay of such an increased attenuation area in or around a hematoma is not yet fully known. For example, etiology of the enhanced ring blush, which appears transiently several days or weeks after the hemorrhage, is still not clearly explained. Figure 7 demonstrates an actual example of such an ambiguous pattern of increased density. With enhancement, a fusiform pattern with increased density appeared

Fig. 5. Case 2. Angiogram of the left carotid artery showing no angiomatous vasculature.

Fig. 6. Case 2. Photomicrograph of the mass, showing a grouping of many arteries, arterialized veins, and venous vessels with thickened walls. Elastica van Gieson, x 19.
along the outer rim of the low-density area. Whether this is a small angiomatous lesion responsible for intracerebral hemorrhage or merely an organized residue in the old hemorrhage was not possible to confirm in this case. In Case 2, an indistinct lesion abutting on the ring blush was surgically explored and a small vascular mass was removed, which was confirmed histologically to be a cryptic AVM.

There is no doubt that CT is the most reliable modality of diagnosis of cryptic vascular malformations, but many of these AVM's can still escape detection on account of their very small size, destruction of the malformation by hemorrhage, and difficulty in interpretation of various patterns of increased attenuation areas in associated hematoma. Non-opacification of a vascular malformation on angiography, which is the main cause of a cerebral angioma being "cryptic," may result from several factors. Stagnant or slow blood flow in the angioma and various degrees of thrombosis in the vessels of the angioma seem to be the main causes. The small size of the angioma and destruction or compression of the angioma by resultant hemorrhage are other contributing factors. The former two factors are satisfactorily solved by CT scanning. Recently, Kramer, et al., reported CT findings in cases of angiographically occult cerebrovascular malformations that were thrombosed.

The latter two factors remain more refractory to CT. According to Mori, et al., the visualization threshold of intracerebral lesions by CT scanners currently used in many hospitals is 0.5 to 1.5 cm. The size of cryptic angiomas varies, with some of them as small as 1 to 2 mm in maximum dimension, and others microscopic in size. Advances in CT technology will produce scanners with higher resolution that will allow such lesions to be diagnosed. In addition, analysis of the pathophysiological background of high-density areas in the aging process of intracerebral hemorrhage and establishment of correlation of CT figures with neuropathological findings in cryptic angiomas of heterogenous histology will undoubtedly contribute to diagnosis of cryptic angioma by CT.

References

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