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Spontaneous Hemorrhage from a Cerebral Venous Malformation Related to Thrombosis of the Central Draining Vein: Demonstration with Angiography and Serial MR

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Summary: A cerebral venous malformation ruptured after acute thrombosis of its central draining venous channel, leading to intracerebral hemorrhage. The CT, MR, and angiographic studies unequivocally demonstrated subsequent progressive recanalization of the malformation, providing proof that venous malformations can spontaneously bleed and that venous thrombosis may be associated with, and perhaps produce, the hemorrhage.

Index terms: Cerebral hemorrhage; Thrombosis, venous; Veins, cerebral

Venous malformations are composed of thin-walled vessels with minimal elastic or smooth muscle layers, separated by normal intervening neural parenchyma. They are most commonly found in the frontal lobes and posterior fossa, usually in a subcortical or periventricular location. Malformed deep medullary veins typically drain into a single large centrally located vein or varix.

Since the introduction of magnetic resonance (MR) imaging, venous malformations of the brain have become the most frequently detected cerebral vascular malformation. Despite this, the incidence of venous malformation–associated hemorrhage remains unclear, with reported incidence ranging from 0% to 43% of cases (1–9). Some authors propose that cavernous malformations, which are frequently associated with venous malformations, are the true source of hemorrhage in most cases of "complicated" (hemorrhagic) venous malformations (5–8). Also, given the relatively high frequency of detecting incidental venous malformations with MR imaging in asymptomatic persons (4), reports indicating a high incidence of associated hemorrhage may be questioned.

We present a case of computed tomography (CT)- and MR-documented acute thrombosis of the main outflow of a venous malformation temporally and spatially associated with parenchymal hemorrhage. Sequential MR evaluation supports venous thrombosis as a mechanism by which this venous malformation bled.

Case Report

A 34-year-old white woman with no significant medical history presented to the emergency room with persistent headache. On physical exam, she had a left homonymous hemianopsia and bilateral papilledema. CT scan at that time revealed a 4×3×5-cm intracerebral hematoma in the right parietotemporal region with mass effect (Fig 1A). More inferiorly, in the posterior temporooccipital region, there was a contiguous funnel shaped 1-cm area of acute blood clot (arrow, Fig 1A), connected to the dominant larger clot by a thinner linear high-density band, and broadest inferiorly near the surface of the tentorium. Angiography performed that day showed mass effect from the dominant clot, but no arterial or venous abnormality (Fig 1B).

An MR study the next day confirmed the presence of the acute intracerebral hematoma in the right parietotemporal region and the smaller adjacent clot inferiorly (arrow, Fig 1C). Coronal images demonstrated a linear band extending inferiorly from the large hematoma, progressively dilating as it approached the upper surface of the tentorium to form a pyramidal structure that contained the smaller clot (Fig 1D). The pyramidal structure appeared to represent the central draining vein of a venous malformation, containing intraluminal thrombus.

The patient was treated conservatively and discharged 9 days after admission. One month later, MR demonstrated peripheral enhancement of the still thrombosed draining vein (Fig 1E), and angiography still did not demonstrate any vascular anomaly. A 2-month follow-up MR

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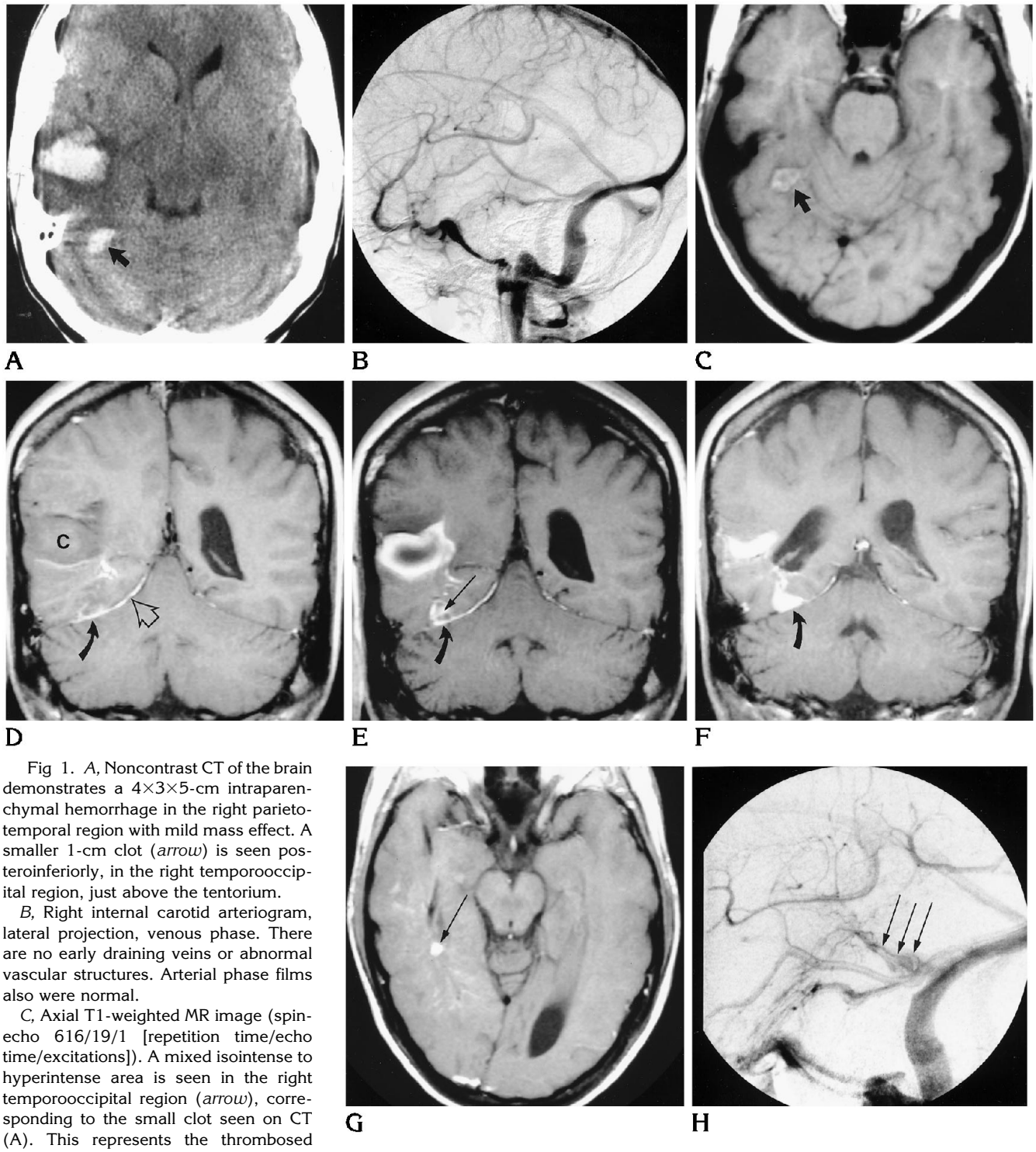

Fig 1. *A,* Noncontrast CT of the brain demonstrates a 4×3×5-cm intraparenchymal hemorrhage in the right parietotemporal region with mild mass effect. A smaller 1-cm clot (*arrow*) is seen posteroinferiorly, in the right temporooccipital region, just above the tentorium.

B, Right internal carotid arteriogram, lateral projection, venous phase. There are no early draining veins or abnormal vascular structures. Arterial phase films also were normal.

C, Axial T1-weighted MR image (spin-echo 616/19/1 [repetition time/echo time/excitations]). A mixed isointense to hyperintense area is seen in the right temporooccipital region (*arrow*), corresponding to the small clot seen on CT (A). This represents the thrombosed central draining varix of the malformation.

D, Coronal postcontrast T1-weighted MR image (spin-echo, 516/19/1). The larger (isointense) parenchymal clot in the right parietotemporal region (*C*) and the thrombosed varix abutting the tentorium in the inferior temporooccipital region (*arrow*) are connected by an enhancing linear band. There is some enhancement of the adjacent tentorium (*open arrow*), probably related to collateral flow draining this region. A T2-weighted study (not shown) revealed marked hypointensity in areas of clot.

E, One-month follow-up coronal postcontrast T1-weighted MR image (spin-echo, 516/19/1). The smaller, pyramid-shaped clot within the thrombosed varix along the tentorium (*arrow*) now demonstrates peripheral enhancement (the wall of the draining vein), whereas the center remains predominantly isointense to hypointense (intraluminal clot). A small focal area of enhancement within the clot (*long arrow*) suggests early recanalization. The larger parenchymal clot now is peripherally hyperintense (same appearance as before infusion). *Continues.*

study showed almost complete recanalization of the draining vein (Fig 1F), as well as an extensive fine network of radially arranged enhancing medullary veins draining into the central recanalized varix, characteristic of venous malformation (Fig 1G). The typical "caput medusae" appearance of dilated medullary veins filling in the normal venous phase also was seen at angiography 3 months after presentation (Fig 1H), with the funnel-shaped varix now seen to drain toward tentorial veins, as previously suspected from the MR appearance.

Discussion

This case clearly demonstrates hemorrhage from a venous malformation. We believe that bleeding was related to an increase in intralesional pressure related to thrombosis of the central draining vein. Lesional thrombosis initially prevented definitive diagnosis, which was delayed until the lesion recanalized.

On the initial CT and T2-weighted MR, the thrombosed draining vein could be clearly seen as a smaller area of blood clot in the posterior temporooccipital region, adjacent to the tentorial surface and distinct from the secondary intracerebral hematoma. Appreciation of the significance of the well-marginated linear clot as a marker for the presence of venous thrombosis led us to consider the diagnosis at an earlier stage than was possible given the lack of direct visibility of the medullary component of the lesion. Enhancement along the tentorium on the initial MR was likely related to collateral flow in tentorial veins, in response to thrombosis of the usual drainage route.

The initial angiogram failed to demonstrate the venous malformation, as would be expected with lesional thrombosis. As the malformation and the draining vein began to recanalize, there was enhancement (on MR) of the fine network of radially arranged veins forming the head, or "caput medusae," of the malformation, and the lesion became angiographically apparent on the final study 3 months after presentation.

There has been and there remains considerable controversy regarding the propensity of venous malformations to bleed spontaneously. The reported incidence of radiographically demonstrable hemorrhage ranges from 0% to as high as 43% (1–9). Lobato reported that 96% of venous malformations have evidence of previous hemorrhage on histologic examination (10). Some authors feel that cavernous malformations or other angiographically occult arteriovenous malformations actually are responsible for hemorrhages previously attributed to venous malformations (5, 6). Cavernous malformations have been found in association with as many as 27% of venous malformations (6). On the other hand, Wilms et al found no associated cavernous malformations in a series of 28 patients with venous malformations studied by both MR and angiography, despite the fact that four of the cases had bled (7). Although pathologic confirmation is lacking in our case, no characteristic features of cavernous malformation were present on any of the studies.

One may question why a group of veins deep within the cerebrum should spontaneously bleed, given that these veins provide the "normal" drainage route for a portion of brain parenchyma. It has been proposed that elevated venous pressure in a venous malformation may lead to hemorrhage directly (1, 5) or indirectly by inciting the development of an associated cavernous malformation that then bleeds (9). Spontaneous thrombosis of venous malformations or acquired venous occlusive disease has been suggested as a possible mechanism for hemorrhage or infarction by a number of authors (1, 5, 9–12). Location and size, once thought to be risk factors, have not been shown to correlate with a bleeding tendency on more recent studies (2, 6). Although it has been argued that venous malformations are low-flow, low-pressure lesions that should not spontaneously bleed, cavernous malformations, which

F, Two-month follow-up coronal postcontrast T1-weighted MR image (spin-echo, 516/19/1). The pyramid-shaped structure that once contained clot has completely recanalized and now enhances uniformly (*arrow*). This is more clearly seen to be the dilated varix of the venous malformation, which drains into the tentorial venous plexus. The uniformly bright parenchymal clot has decreased in volume.

G, Two-month follow-up axial postcontrast T1-weighted MR image (spin-echo 616/19/1). The upper segment of the fully recanalized central draining vein (*long arrow*) is now well demonstrated by uniform enhancement on MR. Numerous fine linear structures radiating outward form the classic "caput medusae" appearance of the medullary venous portion of the venous malformation.

H, Three-month follow-up right internal carotid arteriogram, lateral view, venous phase, demonstrates the completely recanalized venous malformation. The caput medusae appearance of abnormal medullary veins can be seen draining into a dilated, pyramid-shaped varix (*arrows*), which then empties into the tentorial venous plexus. This closely corresponds to the MR findings.

frequently bleed, also have low intralesional pressures and low flow velocities. It is interesting to note that focal stenosis of the draining vein of a venous malformation may be seen at angiography just at the point where the vein penetrates the dura to enter a dural sinus (1), further supporting our theory that venous stenosis and thrombosis may increase the risk of hemorrhage (1).

Our case is clearly a thrombosed venous malformation with associated hemorrhage. Two other similar, but less well-documented, cases have been reported in the literature (11, 12). Late recanalization of acutely thrombosed venous channels might explain why some reported cases of hemorrhagic venous malformation have initially negative angiograms (8). Because clinical management of venous malformations and patient counseling may hinge on the incidence of hemorrhage, it is important to establish the mechanism by which venous malformations may bleed. Although it is still likely that venous malformations are for the most part benign developmental anomalies, one should not dismiss the possibility that they may be the source of significant hemorrhage in some patients and that hemorrhage may be a direct result of spontaneous thrombosis of veins draining the malformation.

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