Mixed arteriovenous malformation and capillary telangiectasia: a rare subset of mixed vascular malformations

Case report

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In this report, the authors discuss the case of a patient with a mixed cerebrovascular malformation in which an arteriovenous malformation (AVM) was associated with a capillary telangiectasia. Recent reports have contained reviews of various subsets of mixed malformations. To the authors’ knowledge, however, this is the first report of a mixed vascular malformation with both arterial and capillary components. The patient underwent complete resection of the AVM after presenting with a clinical hemorrhage. She required a second operation to resect the capillary telangiectasia after new symptoms developed several months following the first procedure. The authors conclude that a mixed AVM-capillary telangiectasia is a rare but distinct entity.

KEY WORDS • arteriovenous malformation • capillary telangiectasia • mixed vascular malformation • pathology

CEREBROVASCULAR malformations have long been classified in the neurosurgical literature on the basis of their vascular architecture and this classification system has become a widely accepted standard. Recently, several authors have identified and classified mixed vascular malformations as a specific subset of vascular malformations. These mixed malformations have a less clearly defined pathogenesis, natural history, and treatment. Furthermore, the various appearances of subtypes of vascular malformations on angiography and magnetic resonance (MR) imaging often make the diagnosis of a mixed malformation less obvious. We report the case of a patient who presented with a clinical hemorrhage that led to the presumptive diagnosis of an arteriovenous malformation (AVM) based on imaging studies. Two surgical resections revealed this lesion to be a mixed AVM and capillary telangiectasia. Although other subtypes of mixed vascular malformations have been well described, the entity of an AVM combined with a capillary telangiectasia has not, to our knowledge, been previously discussed in the neurosurgical literature.

Case Report

This 42-year-old woman presented to an outside institution with a 1-month history of vertigo that worsened with rapid changes in position. During the year prior to this presentation, the patient had noted an increasing frequency of occipital headaches. In January 1993, she had experienced an unusually severe headache associated with nausea, vomiting, and diplopia.

First Examination. The patient’s neurological examination was notable for diminished coordination on the right side of her body with decreased agility at tandem walking and mild dysdiadochokinesia. Her reflexes and motor, sensory, and cranial nerve testing were normal.

Magnetic resonance imaging demonstrated a highly vascular lesion, measuring 3 cm in diameter, that was located in the right posterior inferior cerebellum (Fig. 1 left). No subacute or chronic blood was noted. Cerebral angiograms revealed a 3-cm AVM with a dense venous-phase enhancement, early draining veins, and a parenchymal blush (Fig. 1 center and right). The lesion was supplied primarily by the right posterior inferior cerebellar artery.

First Operation and Pathological Findings. At surgery, a 3-cm AVM nidus was resected and its identity was confirmed by pathological examination (Fig. 2). A postoperative angiogram showed complete resection of the AVM.

First Postoperative Course. Postoperatively, the patient did well, with decreased vertigo and improvement in her diplopia. One month later, the patient began to develop
difficulty with speech, poor fine motor skills of the right hand, and worsening of her vertigo, diplopia, and gait.

Second Examination. A repeated MR image showed extensive, marked enhancement involving the right cerebellar hemisphere that suggested a residual non-AVM type of vascular malformation (Fig. 3 left).

The patient was admitted to Stanford University Medical Center for surgery. Preoperative angiograms showed no arteriovenous shunting or early draining vein (Fig. 3 center and right).

Second Operation and Pathological Findings. A vascular malformation, 5 cm in diameter, was found in the right cerebellar hemisphere, extending to the vermis bilaterally and to the left middle cerebellar peduncle. The vascular malformation also extended deep to the right dentate nucleus and the right middle cerebellar peduncle. Pathological investigation confirmed a capillary telangiectasia, with no elastica noted in the vessel walls (Fig. 4). Intervening cerebellar tissue was noted between the vascular walls. Postoperative MR imaging showed complete resection of the telangiectasia (Fig. 5).

Second Postoperative Course. Postoperatively, the patient exhibited remarkable improvement. Twelve months after surgery she had complete resolution of her dysarthria, vertigo, nausea, vomiting, and diplopia with minimal residual gait ataxia and right upper-extremity ataxia. By 24 months after surgery, the patient had resumed her previous activities.

Discussion

A widely accepted classification system groups vascular malformations into arteriovenous, cavernous, venous, and capillary subtypes. Arteriovenous malformations are described as a confluence of arteries and veins without an intervening capillary bed. Few of these are angiographically occult and most have a characteristic angiographic appearance of arteriovenous shunting and early venous drainage. Capillary telangiectases are composed of capillaries whose walls lack elastic and muscle fibers. The sizes of the vessels vary widely and may appear as cavernous spaces. Capillary telangiectases are angiographically occult, usually asymptomatic, and can be found in 0.04 to 0.1% of autopsy cases.

The concept of a mixed vascular malformation has only recently been addressed in the literature. Recent published reports of large series of vascular malformations have indicated that mixed vascular malformations may have been underreported in previous pathological reviews and that, even once identified, these mixed vascular malformations may have been misclassified. Furthermore, the literature reflects a lack of consistent terminology.

The pathophysiology of mixed vascular malformations is also not clearly understood and it remains questionable whether these hybrids represent transitional forms of vascular malformations, with unique natural histories and hemorrhage rates, or whether their natural history and clinical relevance can be explained simply on the basis of combining the characteristics of the various separate vascular subtypes. To date, only small series of mixed vascular malformations have been reported, precluding a definitive description of the natural history, pathogenesis, and clinical relevance of these lesions. In a recent review
of 280 cases of vascular malformations, 14 mixed malformations were identified.\(^3\) Other case reports have revealed similar hybrid malformations.\(^12,19\)

Most vascular malformations are believed to have a congenital origin.\(^{16,18,20,21,28}\) Theories have focused on the failure of blood vessel involution during development, resulting in formation of a vascular malformation.\(^{26}\) Capillary telangiectases may develop this way because the normal brain has numerous capillary channels that involute as development occurs.\(^7\) Any failure in involution due to an aberrant “developmental signal” may result in a capillary telangiectasia. In recent reports, Mullan, et al.\(^{20,21}\) have supported this view of vascular malformation development. In addition, these authors believe that the hyperemic cortex adjacent to an AVM may be composed of “satellite” vessels captured by the AVM during the postnatal period.\(^{20}\) Takemae and colleagues\(^{31}\) described “modja-modja” vessels, a perinidal hypervascular network associated with large AVMs, noting a similarity between these vessels and abnormal capillaries. However, unlike capillary telangiectases, which are occult, modja-modja vessels are present on angiograms obtained after the AVM has been resected. Chin, et al.,\(^6\) described a group of patients with “diffuse AVMs,” lesions that contain normal cerebral tissue interspersed between abnormal vessels. In contrast to the AVM in our case, such diffuse AVMs do not have a well-defined compact nidus. All of the diffuse AVMs studied by these researchers were angiographically apparent, contrasting with the angiographically occult telangiectatic component of the mixed vascular malformation in this case. Furthermore, diffuse AVMs have the histopathological characteristics of true AVM vessels in the malformation, as opposed to the capillary vessels, which lack elastin, as noted in the present case.

Although capillary telangiectases are believed to be much less aggressive than other vascular malformations and most are incidental autopsy findings, they may change local hemodynamic structures in such a way that a true AVM could develop. Previous researchers have hypothesized that small microhemorrhages may induce angiogenic factors, resulting in new vessel development.\(^{2,23,29}\)

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**Fig. 3.** Left: Follow-up gadolinium-enhanced T\(_1\)-weighted MR image, obtained several months after the first surgery, revealing an extensive marked enhancement of the right cerebellar hemisphere. Center and Right: Angiograms obtained before the second surgery showing no arteriovenous shunting or early draining vein in either the anteroposterior (center) or lateral (right) view.

**Fig. 4.** Photomicrographs showing the capillary telangiectasia. Left: H & E. Original magnification \(\times 100\). Right: Eosin and van Gieson. Original magnification \(\times 250\).
Alternatively, the shunting through a capillary telangiectasia may cause arteriolization of some vessels, leading to formation of an AVM. Others have proposed that hemodynamic stress can cause the thickening and hyalinization of vascular walls. Another hypothesis involves pluripotent cells within vascular walls of capillaries and veins, which may develop into muscular cells with arteriolization.

A case such as the one presented here, which represents the combination of an AVM and a capillary telangiectasia, has not been previously described in the literature. The natural history of AVMs is well known, as is their propensity to bleed. Capillary telangiectases are believed to be less aggressive, although there have been documented cases of clinical hemorrhage. In our patient, the initial clinical hemorrhage was presumed to have been caused by the AVM and clinical improvement was seen after its resection. The cerebellum surrounding the AVM appeared as pink hyperemic tissue; this was believed to be an angiomatic change that accounted for the difference between lesion size on the initial MR image and on the angiogram. In retrospect, this hyperemic region may have harbored the capillary telangiectasia; the appearance of the tissue was similar to other gross appearances of telangiectases. The location of the capillary vascular structure in the middle cerebellar peduncle is consistent with the common location of cerebellar capillary telangiectases found in autopsy series. The enlargement of the capillary telangiectasia between the first and second surgeries may have been caused by a decompression of this structure after the AVM resection, an increase in blood flow through the capillary structure as a result of AVM resection, or growth following stimulation of the vessels by angiogenic factors. The angiogram obtained after the first resection demonstrated complete resection of the AVM; thus the patient’s symptoms prior to the second surgery may have been caused by an increasing mass effect in the posterior fossa as a result of capillary telangiectatic growth, a phenomenon previously reported.

Passive hyperemia caused by a venous obstruction associated with the AVM does not seem likely because this would have been expected to resolve after resection of the true AVM with elimination of the arteriovenous shunting. Resection of the capillary telangiectasia resulted in a significant improvement in the patient’s symptoms.

The radiographic appearance of capillary telangiectases is well documented. They are generally occult on angiograms and on MR imaging, although they can appear on MR imaging as areas of decreased signal intensity. Often, they are too small to be adequately imaged on computerized tomography or MR imaging. If there is hemorrhage from the capillary telangiectasia, MR images may show evidence of a signal change caused by subacute or chronic blood, depending on the age of the lesion. In our case, an angiogram obtained before resection of the capillary telangiectasia revealed no evidence of arteriovenous shunting or early draining vein, which is consistent with an occult malformation. Magnetic resonance imaging performed after the AVM resection but before the telangiectasia resection revealed an increased signal on T1-weighted imaging; this signal enhanced after administration of gadolinium. Although this is not the classic appearance of a telangiectasia, the large size of this telangiectasia, as well as the fact that there had been previous surgery in the area, may have contributed to the signal change.

In summary, we conclude that a mixed vascular malformation with AVM and capillary telangiectasia exists as a clearly defined entity. The lack of previous reports on a mixed AVM and capillary telangiectasia may be due to an unclear understanding of these hybrid malformations from both the neurosurgical and pathological point of view. A closer evaluation of intracranial vascular malformations may indicate that mixed malformations and, specifically, mixed AVMs and capillary telangiectases are more common than previously believed.

References

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