Postnatal growth and development of a cerebral arteriovenous malformation on serial magnetic resonance imaging in a child with hemangiomatosis

Case report

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✓The authors report the case of a 3-week-old girl with two enhancing extraaxial masses in the posterior fossa, one in the left cerebellopontine angle (CPA) and the other to the right of the vein of Galen. Serial magnetic resonance images obtained in this patient at 3 months and then at 2.5 years of age documented regression of the enhancing mass in the left CPA and development of a cerebellar brain arteriovenous malformation (AVM) in the same CPA location. Also documented were regression of the pineal region mass and formation of the major draining vein of the AVM. The findings in this case support the theory that cerebral AVMs have early postnatal growth potential.

KEY WORDS • arteriovenous malformation • hemangioma • pediatric neurosurgery

It is believed that the origin of most AVMs is congenital, with embryological errors occurring in capillary or vein morphogenesis.4,11,12,20 Nevertheless, there is no strong evidence regarding the sequence and timing of events from vascular maldevelopment to cerebral AVM formation. A few rare cases have been reported of de novo acquired cerebral AVM development later in life, and most of these lesions are small, with microshunting, and are hypothesized to arise in association with local reactive angiogenesis in previously injured parenchyma.4,13 These lesions are probably different, however, from the nidal and fistulous cerebral AVMs that are typically diagnosed in clinical practice. We report the case of a child with an infratentorial cerebral AVM that developed postnatally either adjacent to or from an enhancing soft-tissue mass; the lesion was documented using serial cross-sectional MR imaging.

Case Report

This patient was born with cardiomegaly and congestive heart failure. A diagnosis of umbilical cutaneous hemangioma and multiple hepatic hemangiomas was made, and she underwent embolizations of her hepatic hemangiomas, which effectively treated her congestive heart failure. An MR image of the brain obtained when the patient was 3 weeks old revealed ventriculomegaly and enhancing masses in the pineal region and left CPA (Fig. 1). Based on these findings, she underwent a 13-month steroid treatment course. At 2.5 years of age, she was slightly developmentally delayed, had just begun walking with a hesitant gait, and had an intermittent downward gaze. Cardiac evaluation showed mild ventriculomegaly with normal function. Follow-up MR images obtained at this time showed less ventriculomegaly and regression of the pineal region mass and formation of the major draining vein of the AVM (Fig. 2).

Because of the development of prominent vessels, including deep veins, despite involution of the enhancing mass, conventional cerebral angiography was performed, the results of which confirmed the suspicion of a cerebral AVM (Fig. 3). One year later, MR imaging did not demonstrate vessel regression, but rather increased size of the draining veins of the AVM and persistent flow voids within the cerebellum. Two brain AVM embolizations were performed with glue (N-butyl cyanoacrylate), one when the patient was 3 and the other when she was 4 years of age, in preparation for future radiosurgery. At the age of 5 years, her MR images
demonstrated no new changes, and she had chin-up and left-sided face-turn posture as a result of her restricted upward gaze, but results of her eye examination were otherwise normal; neurologically she was stable, aside from persistent mild developmental delay. At the age of 6 years she experienced an acute headache, lost consciousness, and was immediately transported to a local hospital, where she died of a cerebral hemorrhage.

Discussion

Intracranial capillary hemangiomas are rare, with fewer than 10 reported cases in the literature.\(^1\),\(^6\),\(^17\),\(^19\) In these reports, the lesions were shown to be isointense or slightly hyperintense on T\(_1\)-weighted and hyperintense on T\(_2\)-weighted MR images, and uniformly densely enhancing after contrast administration. Pathologically, intracranial capillary hemangiomas show a lobular proliferation of closely packed, thin-walled capillaries.\(^6\) The main differential is hemangioblastoma, which is more often cystic, with an enhancing nodule, and is more likely to present with hemorrhage.\(^7\) In the two reported cases of multiple intracranial capillary hemangiomas, as in our patient, the lesions regressed with nonsurgical management.\(^1\),\(^7\)

Cerebral AVMs are thought to be congenital vascular lesions;\(^4\),\(^11\),\(^12\),\(^20\) however, the mechanism, sequence, and timing of development of cerebral AVMs have not been elucidated, only theorized. In this case of what we believe to have been rare intracranial involuting capillary hemangiomas,\(^1\),\(^6\),\(^17\),\(^19\) serial MR imaging revealed relatively rapid postnatal development (within 2 years) of either the transformation of a hemangioma into a cerebral AVM or the growth and development of an adjacent cerebral AVM.

The first possibility seems unlikely because capillary hemangiomas, although rare intracranially, are relatively common in other parts of the body, and despite this greater prevalence elsewhere, transformation from capillary hemangioma to true AVM has not been reported. Capillary hemangiomas or other vascular tumors can mimic an AVM on angiographic studies by demonstrating arteriovenous shunting, but this occurs within an enhancing mass, as shown on MR cross-sectional imaging.\(^3\),\(^9\),\(^14\) Arteriovenous shunting in capillary hemangiomas regresses as the mass involutes.\(^2\) In our case, follow-up MR imaging demonstrated no residual enhancing mass containing the flow voids (Fig. 2). The few intracranial capillary hemangiomas that have been evaluated angiographically demonstrated sharp margins with intense persistent staining in a lobular pattern, and slight enlargement of feeding arteries without the appearance of a cerebral AVM.\(^1\),\(^6\),\(^18\)

The second, and what we believe to be the more likely possibility, is the growth and development of an adjacent and coincidental cerebral AVM. We suspect that this child’s cerebral AVM was a result of congenital vascular maldevelopment during embryogenesis, but that its growth and de-

![Fig. 1. Serial MR images obtained when the patient was 3 weeks old. Axial T\(_1\)-weighted MR image obtained without addition of contrast material (A), and axial (B) and sagittal (C) T\(_2\)-weighted images obtained after addition of contrast material demonstrating well-circumscribed, isointense, densely enhancing masses in the left CPA and right pineal region. The lesions were hyperintense on T\(_1\)-weighted images (not shown). A prominent flow void representing a vessel is present adjacent to or within the lateral margin of the left CPA mass. Ventriculomegaly and dilated cerebrospinal fluid spaces are present. A T\(_1\)-weighted axial MR image of the liver (D) demonstrates numerous hepatic capillary hemangiomas.](image1)

![Fig. 2. Follow-up MR images of the patient obtained when she was 2.5 years of age. Axial T\(_1\)-weighted (A) and T\(_2\)-weighted (B) MR images of the brain obtained without addition of contrast material, and coronal (C) and sagittal (D) T\(_1\)-weighted images of the brain obtained after delivery of contrast agent demonstrating regression of the well-circumscribed, isointense on T\(_1\)-weighted, hyperintense on T\(_2\)-weighted, densely enhancing masses in the left CPA and right pineal region. In the left CPA, however, numerous and enlarged flow voids are present with no soft-tissue component, and a prominent vein is demonstrated in the pineal region. Prominent flow voids are present within the cerebellar parenchyma.](image2)
Development occurred rapidly in the early years of life rather than in utero. The postnatal growth and development of the child’s cerebral AVM, as demonstrated on serial MR imaging studies, was disproportionately more than that of adjacent normal brain vessels. The influence of the presence of an adjacent involuting hemangioma and of the systemic steroid treatment in affecting this development is unknown.

The patient’s ventriculomegaly and dilated cortical subarachnoid spaces were evidence of dysfunction in cerebrospinal fluid absorption and probable venous hypertension. In an animal model, venous hypertension was associated with dural AVM development; however, the role of venous hypertension in cerebral AVM development is unknown. The timing of the lesion’s development may involve genetic influences, with or without an association with neurocutaneous diseases, local and systemic factors, and apoptosis.

If a general theory of cerebral AVM origin and development is proposed, the observations in our case should be explained. Mullan and colleagues speculated that cerebral AVMs are related to an occurrence at the stage of absorption of multiple pial-to-dural subarachnoid veins, at the point in development at which the fetus reaches a 40- to 80-mm length. Mullan and associates suggested that cerebral AVMs, in contradistinction to vein of Galen malformations, develop after birth because even with the availability of intrathecal and neonatal ultrasonography, investigators had not uncovered an equivalent number of cerebral AVMs despite the higher prevalence of these lesions. These authors surmised that cerebral AVMs had postnatal growth potential and were mostly undetected at birth, but that the lesions were already in place before birth. Our observations in this case are consistent with this theory.

Conclusions

Using serial MR imaging, we observed early and relatively rapid postnatal growth and development of a cerebral AVM in a child with hemangiomatosis. The findings in this case support the theory that cerebral AVMs have postnatal growth potential.

References


Fig. 3. Conventional cerebral angiograms, anterior (upper) and lateral (lower) projections, confirming the diagnosis of brain AVM. Angiographic studies demonstrate a large left superior cerebellar hemispheric AVM supplied by the following feeding arteries: the left superior cerebellar artery, left posterior inferior cerebellar artery, and left anterior inferior cerebellar artery. The venous drainage of the malformation flows mainly into the dilated left lateral mesencephalic vein, the vein of Galen, and straight sinus.
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