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Spontaneous Regression of Cerebral Arteriovenous Malformation in Hereditary Hemorrhagic Telangiectasia

Harry J. Cloft

Summary: Cerebral arteriovenous malformations (AVMs) are associated with hereditary hemorrhagic telangiectasia (HHT). I report a case of a patient with HHT with four cerebral AVMs in whom one AVM spontaneously regressed during a 5-year period. The spontaneous regression of this AVM in this patient with HHT supports the theory that AVMs associated with this syndrome have a different natural history than that of typical sporadic AVMs.

Hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome, is a disorder with an autosomal dominant inheritance pattern that is characterized by mucocutaneous telangiectatic lesions of the skin, mucous membranes, and viscera and by arteriovenous fistulae in the brain, lungs, and gastrointestinal tract (1). Multiple cerebral arteriovenous malformations (AVMs) are associated with HHT (2–4). Cerebral AVMs in patients with HHT are typically small and have a single arterial feeding pedicle and a single draining vein (2, 4). I report a single case of the spontaneous regression of an AVM in a patient with HHT with four cerebral AVMs.

Case Report

A 15-year-old male patient presented with a ruptured left superior hypophyseal arterial aneurysm. Angiography performed to evaluate the aneurysm demonstrated three supratentorial AVMs that were each smaller than 5 millimeters in diameter and an AVM in the left cerebellar hemisphere that was 10 mm in diameter.

Five years later, the patient underwent angiographic follow-up of the AVMs. Angiography revealed resolution of the AVM in the cerebellar hemisphere and no change in the appearance of the supratentorial AVMs (Fig 1).

Discussion

I report a case of a patient with HHT with multiple cerebral AVMs, with one AVM that spontaneously resolved. Although patients with HHT are prone to having multiple cerebral AVMs (2, 4), to my knowledge, the spontaneous resolution of an AVM associated with HHT has not been described.

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Spontaneous regression of sporadic cerebral AVMs is rare, occurring in about 1% of all cases (5, 6). Like the AVMs in this patient with HHT, sporadic AVMs that undergo spontaneous obliteration tend to be small, have few feeding arteries, and have a single draining vein (5, 6). Unlike this patient with HHT, however, 80–90% of patients in whom sporadic AVMs are spontaneously obliterated present with hemorrhage. The increased local pressure and inflammation caused by hemorrhage may lead to the obliteration of a small AVM. I have no evidence of hemorrhage leading to AVM obliteration in this patient, so we must consider another mechanism of obliteration specific to patients with HHT.

Cerebral AVMs in patients with HHT tend to have a more benign natural history than that of sporadic AVMs, with a bleeding risk of 0.36–0.56% per year (7). Additionally, patients with HHT who have an intracranial hemorrhage due to an AVM tend to have good functional outcomes (8). HHT is associated with three different genetic mutations. Some patients have a mutation of the endoglin gene, whereas others have a mutation of the ALK-1 gene, both of which are active in the transforming growth factor-beta (TGF- β) receptor signaling system (9). A third group of patients does not have mutations of the endoglin or ALK-1 genes; these patients seem to have a mutation of a gene that has yet to be identified (9). The mechanisms by which these mutations lead to AVM malformation are not yet understood. This case also suggests that one or all of these mutations may also lead to formation of an unstable AVM that is prone to spontaneous obliteration.

The spontaneous regression of a brain AVM in a patient with HHT supports the argument that cerebral AVMs in these patients have a natural history different from that of sporadic cerebral AVMs. I report only one case and look forward to further confirmation of this phenomenon in additional cases.

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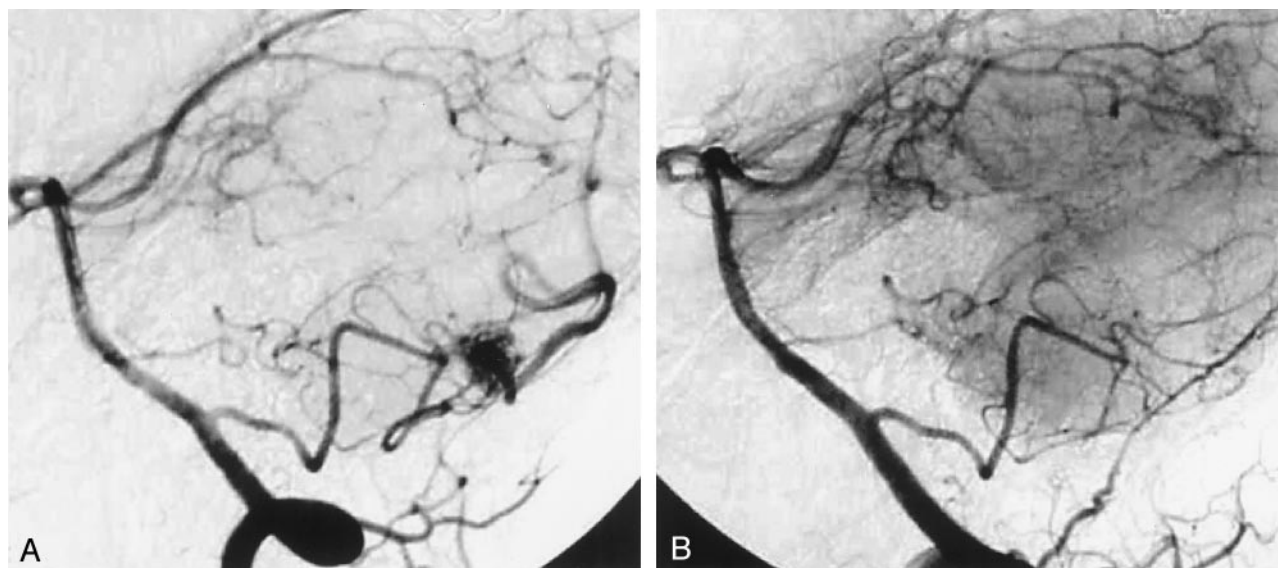


FIG 1. Lateral projection angiograms.

- A, An AVM is fed by the posterior inferior cerebellar artery.
 B, Five years later, spontaneous resolution of the AVM has occurred.

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