Spontaneous disappearance of a cerebral arteriovenous malformation in infancy

Case report

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The authors report a case of the disappearance of an arteriovenous malformation in infancy, demonstrated by follow-up angiography performed 7 months after the original angiograms. Some possible mechanisms whereby a cerebral arteriovenous anomaly is thrombosed are discussed.

KEY WORDS arteriovenous malformation

The spontaneous regression of a cerebral arteriovenous malformation (AVM) is an uncommon occurrence, whether complete or partial. We have found only 13 such instances in the literature, five cases with total disappearance and eight with partial regression. There has been no report of the spontaneous regression of an AVM observed in infancy.

We are reporting an infratentorial arteriovenous anomaly in a 4-month-old baby. Total regression of the AVM was demonstrated in a follow-up study performed 7 months after the original angiograms.

Case Report

This 4-month-old baby girl was admitted on January 18, 1975, because of enlarged head and impaired development. She was born at term, weighing 2800 gm, and had a head circumference of 31 cm. By the age of 9 weeks her head was fairly controllable, but then it became unstable and enlarged in size.

Examination. On admission the patient weighed 5700 gm and her head circumference was 45 cm. The anterior fontanel was 5 × 5 cm in diameter and bulged somewhat. She had increased venous dilatation on the scalp, and sunset phenomenon of the eyeballs. A cranial bruit was audible in both retroauricular regions. The cranial nerves were intact and she could move her extremities freely and equally. There was no abnormal finding in heart sound and cardiac rhythm. An electrocardiogram was interpreted as within normal limits and plain chest film disclosed no cardiac enlargement. No polycythemia was detected in blood tests.

Lumbar puncture revealed a pressure of 350 mm H₂O. The cerebrospinal fluid (CSF) contained 2 lymphocytes/cu mm, and a total protein of 350 mg/100 ml. Indigocarmine (1 ml) introduced into a lateral ventricle did not appear in CSF taken from the lumbar space during a 20-minute dye test.

Right transbrachial angiography on January 24, and left vertebral angiography on January 28, revealed a massive AVM in the posterior fossa fed by the right posterior inferior cerebellar artery, left superior cerebellar artery, and left posterior cerebral
artery. It drained into the straight sinus and right transverse sinus (Fig. 1). Marked internal hydrocephalus was also noticed on the angiograms.

Operation. On January 30, 1975, a ventriculoperitoneostomy was performed. Postoperatively the baby had repeated generalized convulsions. Three days postoperatively an angiographic study was performed in which an avascular area in the right parietotemporal region was revealed, and a postoperative subdural hematoma 1.5 cm thick was evacuated. Right transbrachial angiography performed 25 days later showed no avascular area and the AVM appeared to be slightly smaller.
Spontaneous regression of AVM

Postoperative Course. The patient was discharged on March 4, 1975. After discharge the patient had recurrent attacks of head nodding and electroencephalography showed typical hypsarrhythmia. The attack was fairly controllable with the administration of nitrazepam, diphenylhydantoin, and phenobarbital.

The patient was readmitted on September 9, 1975, for a follow-up study at age 1 year. She weighed 7500 gm, and her head circumference was 44 cm. The anterior fontanel was 2 × 2 cm and not tense. Venous dilatation of the scalp was absent. No cranial bruit nor sunset phenomenon of the eyeballs were noted. Left transbrachial angiography on

Fig. 2. Anteroposterior (left) and lateral (right) views of a right transbrachial angiogram on September 22, 1975 (upper), and a left transbrachial angiogram on September 9, 1975 (lower). The arteriovenous malformation cannot be seen on either angiogram.
September 9, and right transbrachial angiography on September 22, showed that the AVM was no longer present. Furthermore, the right posterior inferior cerebellar artery was not visualized, and the left superior cerebellar artery was smaller in diameter than in the previous angiogram. The major supratentorial arteries, such as the anterior and middle cerebral arteries, were more clearly demonstrated than in the previous studies (Fig. 2). There was no abnormal finding in hemostatic tests and other blood surveys. The patient was discharged on September 23, 1975, and followed in the outpatient clinic. One month later, the attacks of head nodding became well controlled.

Discussion

Various suggestions have been made about possible mechanisms whereby a cerebral arteriovenous anomaly is thrombosed. Svien and Peserico suggested that abnormal vessels in AVM's might have a propensity for thrombosis greater than that of adjacent normal vessels. They postulated that intracranial bleeding, in particular intracerebral bleeding, is the factor resulting in thrombosis of these abnormal vessels. Previous intracranial bleeding has also been noted as an important factor influencing regression of AVM's.

Alteration in cerebral circulation and radiation therapy may influence the regression of the anomalies. Sukoff, et al., reported subtotal occlusion of a massive AVM associated with spontaneous occlusion of the middle cerebral artery. Krayenbühl described two cases in which the arteriovenous anomaly either disappeared or was greatly reduced after carotid ligation. In his second case, carotid ligation was combined with x-ray therapy. In the case reported by Svien and Peserico, regression of the AVM followed a course of radiation therapy.

Pool and Potts remarked that AVM's may undergo thrombosis in the presence of advanced arteriosclerosis. Banker reviewed 48 autopsied cases of occlusive vascular disease affecting the central nervous system in infancy and childhood, and stated that congenital heart diseases, and systemic and focal infections were the most frequent causes of such thrombosis. Since our patient had neither heart failure, infection, nor arteriosclerosis, the disappearance of the vascular anomaly must be considered spontaneous. It might have been assumed, otherwise, that the postoperative subdural hematoma following shunt operation could initiate thrombosis of the AVM.

This is the first known report of total regression of an AVM situated in the posterior fossa in infancy. Cerebral AVM's in infancy have rarely been detected by angiographic study. As reported in the cooperative study on subarachnoid hemorrhage, only 1% of all AVM's are seen in patients below the age of 2 years. In our patient the disappearance of the vascular anomaly, noticed 7 months after the original angiograms, occurred earlier than the 2 to 32 years previously described for the regression of AVM's in older patients. It is well accepted that cutaneous capillary angiomas frequently undergo spontaneous regression in infancy and childhood. It might be postulated that some intracranial vascular anomalies in infancy and childhood might also regress spontaneously.

References

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