MEDICAL MEMORANDA

Cerebral Arteriovenous Malformation Causing Cardiac Enlargement and Epilepsy: Correction after Operation

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The case of a 6-year-old boy with epilepsy and cardiomegaly due to a cerebral arteriovenous malformation is reported. After excision of the malformation the cardiac output fell from 7 l./min., and the heart quickly resumed a normal size.

Case Report

A 6-year-old boy was born full term at home. He had three healthy older siblings. When 3 days old he was admitted to hospital for a month with some obscure heart trouble for which he was given oxygen but not digitalis or diuretics. Subsequently he progressed well with normal milestones. At 18 months he had a series of minor convulsions which were apparently controlled with phenobarbitone. Then at 5½ years he had three right-sided convulsions after he had been off anticonvulsants for two years.

Examination in July 1972, some six months later, showed a healthy active boy with enlarged veins over his scalp and forehead, and particularly in the eyelids and right sclera. The retinal veins were also dilated. A harsh bruit was present in his head, maximal in the left temporal region. The pulse rate was 110, with an increased pulse pressure (B.P. 120/70 mm Hg), and a forceful apex beat which was displaced just beyond the mid-clavicular line. The first heart sound was loud but no murmurs were audible in the heart or neck. X-ray pictures of the chest showed an enlarged heart (fig. 1). An E.E.G. showed high-voltage slow-wave activity at 1 Hz in the left occipital region.

Skull x-ray pictures showed patchy thinning of the skull vault in the left posterior temporal region, while a left carotid and also a vertebral arteriogram performed under general anaesthesia showed a large arteriovenous malformation situated in this region (fig. 2). It shunted practically all the blood carried by the left carotid and the two vertebral arteries directly into the intracranial venous sinuses. The left posterior cerebral artery was greatly enlarged and was the major supply to its deep aspect. The left middle cerebral artery terminated in two large branches which supplied it on the convexity. The left anterior cerebral artery did not fill, presumably as it was being supplied from the opposite carotid artery. The malformation then discharged directly into the left transverse sinus with such power that arterialized blood was forced through the torcula retrogradely into the opposite transverse sinus, and even upwards into Labbé's vein on the opposite hemisphere. The malformation also discharged into the cavernous sinus via basal veins around the brain stem, and into some veins between the

cerebral hemispheres, one of which passed forwards to the left orbit. The arterialized blood then returned to the heart via the two internal jugular veins.

An E.C.G. showed a right bundle-branch block but no evidence of gross ventricular hypertrophy. Echocardiography showed a cardiac output of 7 l./min, which for a boy of 22 kg was more than twice the normal. The end-diastolic volume was 75 ml, the end-systolic volume was 10 ml, and thus the stroke volume was 65 ml. The heart rate was 110.

TREATMENT AND PROGRESS

At operation the malformation was dissected out completely after the feeding arteries had been occluded. Blood replacement was started early to correct a central venous pressure which fell as the feeding arteries were occluded from 8 cm to zero. The child made a speedy and straightforward recovery. The bruit disappeared completely from his head, and the veins in his scalp and retinae were no longer enlarged. The visual acuity remained normal at J1 in each eye, but by stealth a right homonymous hemianopia could be detected. A postoperative E.E.G. was now nearly normal. The heart size became normal (fig. 1).

A left carotid arteriogram repeated two weeks later showed a normal circulation through normal-sized left anterior and middle cerebral arteries with clips placed at the ends of the divided middle and posterior cerebral arteries (fig. 3). The E.C.G. was unchanged. Echocardiography showed that the cardiac output had fallen to a normal level of 3 l./min. The end-diastolic volume was 44 ml, end-systolic volume 11 ml, stroke volume 33 ml, and heart rate 90/min. The blood pressure was 110/80 mm Hg, and so the pulse pressure had become more normal.

He went home on phenytoin 50 mg twice daily, and six months later was reported fit and well.

Comment

Arteriovenous malformations of the brain usually manifest themselves by causing either intracerebral haemorrhage or epilepsy, but rarely by causing cardiomegaly, except in infants. We have been able to find in the literature only a few cases comparable to our own.

Matson (1969) reproduced the arteriograms of a comparable case with a loud bruit, headache, and evidence of cerebrovascular insufficiency. Levine *et al.* (1962) reported four infants with congenital arteriovenous fistulae involving the vein of Galen (a different entity from that which we are describing but similar in that it involved a massive arteriovenous shunt), all of whom at one period of life showed cardiac enlargement and failure. The case most like ours was reported by Taylor *et al.* (1971). This was a 4-year-old boy in whom right carotid arteriography showed a large arteriovenous malformation in the right sylvian region. The heart was enlarged and there was incipient cardiac failure. The scalp veins were engorged. Postoperatively the heart shadow became normal.

In the series of Levine *et al.* (1962) the cardiac output in case 4 was measured by catheterization before operation and four weeks afterwards, and fell from 2.2 to 1.1 l./min. However, there was evidence of later recurrence of the fistula, and the heart shadow remained enlarged.

The method of measurement by echocardiography used in our cases, although needing considerable skill, does not involve needling and is painless (Popp and Harrison, 1970). It therefore deserves a more extended trial, for most advances in therapeutics depend on some degree of measurement before they can be established.

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FIG. 1—X-ray appearances of chest A, before operation showing enlarged heart; and, B, after operation showing normal heart. FIG. 2—Preoperative left carotid arteriogram (early stage). A, Lateral view showing enlarged left posterior cerebral artery and two branches of left middle cerebral artery supplying arteriovenous malformation draining into transverse sinus. B, Anteroposterior view showing internal carotid artery dividing into left posterior cerebral artery (medial) and left middle cerebral artery (lateral) to supply arteriovenous malformation. FIG. 3—Postoperative left carotid arteriogram (comparable stage). A, Lateral view. Internal carotid artery now supplies normal-sized anterior and middle cerebral arteries. Posterior cerebral artery fills faintly. There is no sign of the arteriovenous malformation. B, Anteroposterior view. Basilar artery and both posterior cerebral arteries are well shown filling via posterior communicating artery from internal carotid artery. Left anterior and left middle cerebral arteries are faintly shown in their normal positions.

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References

- Levine, O. B., Jameson, A. G., Nellhaus, G., and Gold, A. P. (1962). *Paediatrics*, 30, 563.
- Matson, D. D. (1969). *Neurosurgery of Infancy and Childhood*, 2nd ed., p. 749. Springfield, Illinois, Thomas.
- Popp, R. L., and Harrison, D. C. (1970). *Circulation*, 41, 495.
- Taylor, J. R., Bell, W. E., and Graf, C. S. (1972). *Journal of Neurosurgery*, 34, 818.