

CHAPTER

16

Strokes in Children

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In children, a sudden neurological deficit, or “stroke” may be caused by hemorrhage, vascular insufficiency, or by neither. The variety of causes is greater than in adulthood. With the one exception of arteriosclerotic cardiovascular disease, a child is subject to all the adult problems, plus a great number peculiar to the young. For example, certain congenital anomalies are of such a nature that they must cause trouble early in life: metabolic defects such as hemophilia and homocystinuria; anatomic anomalies such as those of the heart and great vessels. Bleeding from aneurysms and arteriovenous malformations (AVMs) may occur even in infancy. Furthermore, some diseases which are common to childhood may affect the blood vessels or the blood clotting mechanism, as does leukemia.

To illustrate some of the many problems, nine cases from the Boston Children’s Hospital will be presented, not under any diagnostic heading, but as they presented themselves. Each child had suffered a sudden neurological deficit which suggested a cerebrovascular accident. Some attacks were transient, others progressive. We will let you be the diagnostician, as each story unfolds.

In trying to sort out the diagnostic clues, a neurosurgeon may find himself facing one of the problems of his veterinarian colleague—the patient is not always a good historian and in fact may not speak at all!

CASE HISTORIES

Case 1. Sudden hemiplegia and coma. This 10-year-old boy had been playing at school, walked up two flights of stairs, sat down at his desk, and began to cry because he could not move his right arm or leg. He vomited and became unconscious. After hospitalization, he gradually awakened, but the hemiplegia persisted. A hemihypalgesia was also noted as he became more lucid. Although right-handed, he was not aphasic. Skull films were normal. A lumbar puncture showed grossly bloody fluid. A left carotid arteriogram (Fig. 16.1) showed only slight displacement of midline vessels to the right. There were no occlusions and no recognizable sources of bleeding. He remained stable. The next day a pneumoencephlogram was done, which showed downward displacement of the posterior portion of the left lateral ventricle (Fig. 16.2) and suggested a parietal mass (Fig. 16.3). Since this occurred in 1956, some of today’s diagnostic tests were unavailable. Some centers in Boston were just

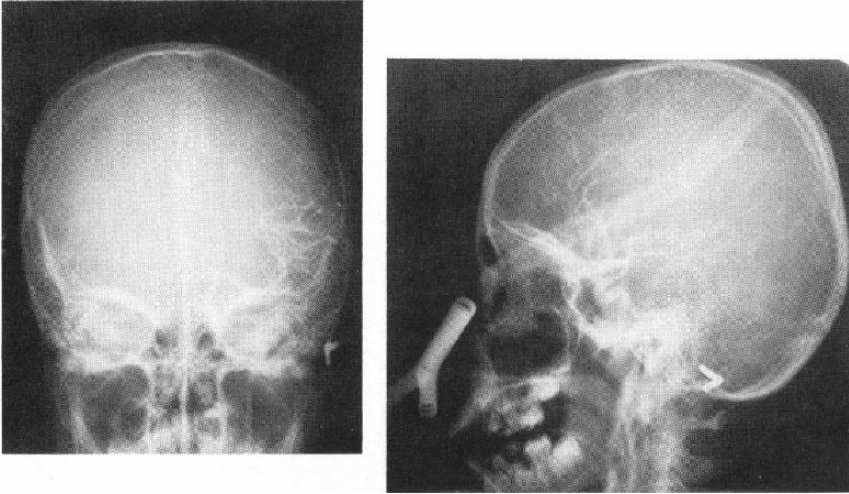


FIG. 16.1. Case 1. Left carotid arteriogram, showing subtle evidence of a parietal mass, but no abnormal vasculature.

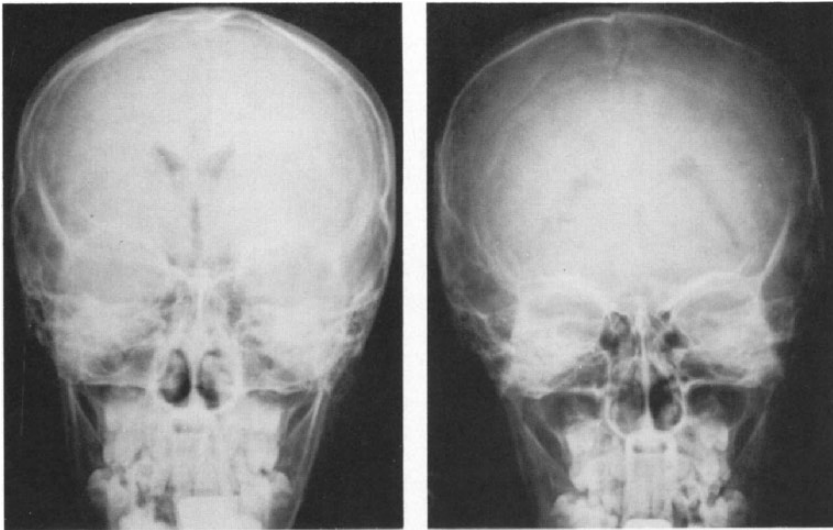


FIG. 16.2. Case 1. Pneumoencephalography confirms presence of left parietal mass.

beginning to scan with radioactive arsenic. The Electronic Musical Instrument Company was not then making computers.

Because the studies indicated a mass lesion, the boy was explored. On the surface of the left parietal area was a small vascular lesion 1.5 cm. in diameter which appeared to contain clot. There was some staining of the arachnoid nearby but no surface clot nor widespread subarachnoid hemorrhage. Careful dissection around this mass revealed that it was

supplied by three small arteries and about the same number of veins which drained toward the midline. These were not thrombosed and were seen to contain arterial blood. After tying, clipping, and dividing the feeding vessels, the lesion literally fell out into the operator's hand (Fig. 16.4). Beneath it within the parietal lobe was a clot estimated at 50 ml. This was removed through two small cortical incisions. The boy recovered the use of his right side but still had a severe seizure problem when last seen 11 years later.

Diagnosis: Thrombosed AVM with intracortical clot.

Here a mass and lack of diagnosis prompted exploration. At operation there appeared to be still a trickle of blood through the malformation, although it did not show on the arteriogram. More often lesions of this size are angiographically demonstrable. Not only may they hemorrhage but they may come to attention in a multitude of ways: headache, dilated scalp veins, asymptomatic bruits (which the pediatrician then wishes he had never heard!), slowly progressive neurological deficit, seizures, or hydrocephalus. They can mimic brain tumors and even multiple sclerosis (1).

Once discovered, what to do with them? Our understanding of the natural history of these malformations is at best inaccurate for we do not



FIG. 16.3. Case 1. Pneumoencephalography showing distortion of body of lateral ventricle.

know how many undiscovered lesions exist. Variation in their size, pattern, and location is infinite. We find it hard to predict for patient or parents which malformation will ever bleed not to mention which will bleed again. The size of the malformation does not correlate with its propensity to hemorrhage. Massive lesions which involve almost an entire hemicranium may not produce symptoms of any kind until adulthood. Microscopic arteriovenous malformations found in surgically evacuated intracortical clots are not even apparent on preoperative arteriograms.

In general, if you can obliterate the shunt without hurting the patient, do so. The likelihood of doing some harm and the nature and extent of that deficit which the patient might have to exchange for eradication can only be decided on an individual basis. Certainly prophylactic ablation is desirable if the price to be paid is nonexistent or small, for a child has a long life ahead and the possibility of getting into trouble is greater than when such a malformation is discovered in middle age.

Successful treatment of these lesions necessitates interruption of the shunt or shunts. Ideally, this should be done at the point or points of entry of arterial feeders into the venous system. Ligation of the arterial feeders far proximal to the site of the shunt must ultimately fail unless there happens to be no collateral channel between the site of ligation and

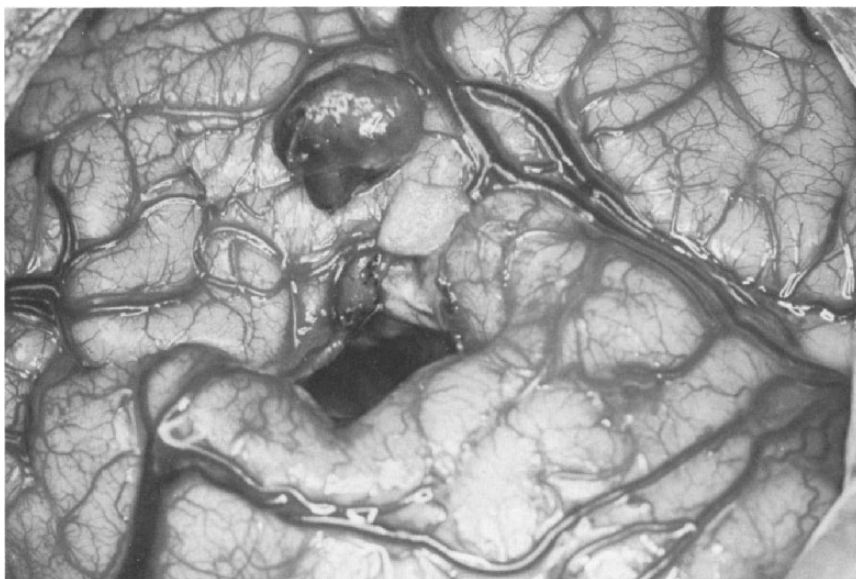


FIG. 16.4. Case 1. Operative photograph of small superficial AVM and cavity in parietal lobe from which clot was evacuated.

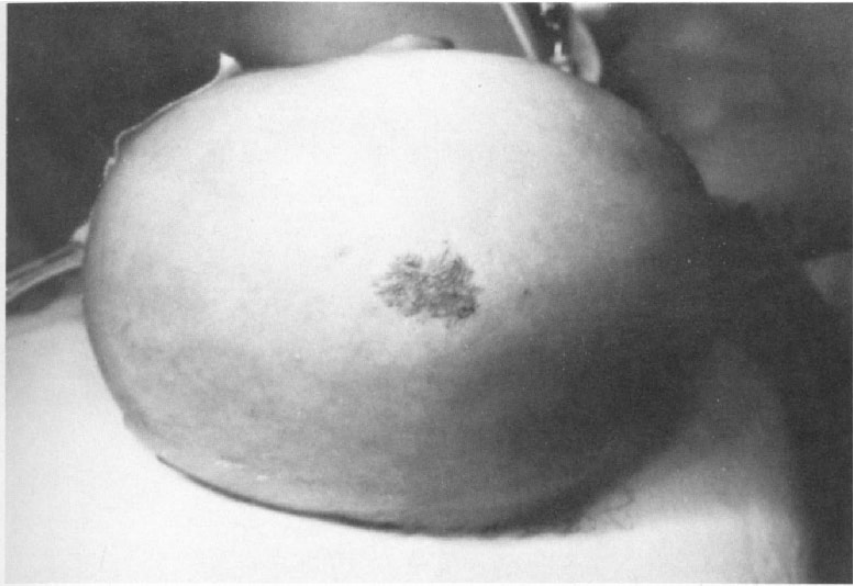


FIG. 16.5. Angioma of scalp of 5½-week-old girl with large AVM of right parietal lobe.

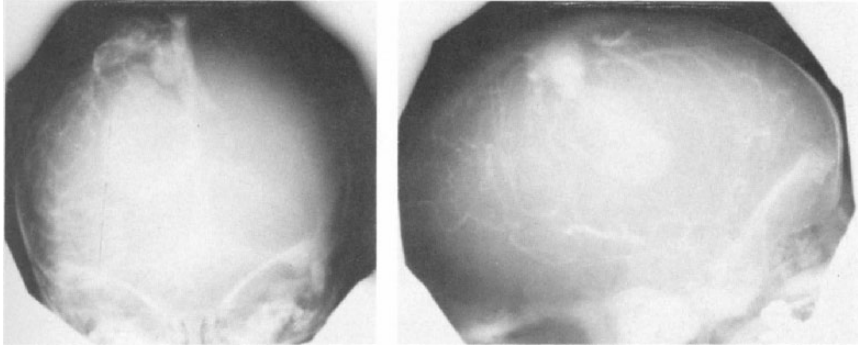


FIG. 16.6. Right carotid angiogram of patient shown in Figure 16.5; AVM appears tri-lobed. Deep large lobe fills and drains from above.

the lesion. This is rarely the case. Ideally, a tie or clip must be placed exactly at the lesion, as, for example, in controlling primary malformations of the vein of Galen (3). The same principle applies to embolization techniques: occlude the shunt without occluding an important artery and without making the occlusion too proximal. Surgery, embolization, or a combination of both, aided by the operating microscope and all the instrumentation that it has brought, are our best tools at the moment.

Some lesions are awesome but manageable. A 5½-week-old girl

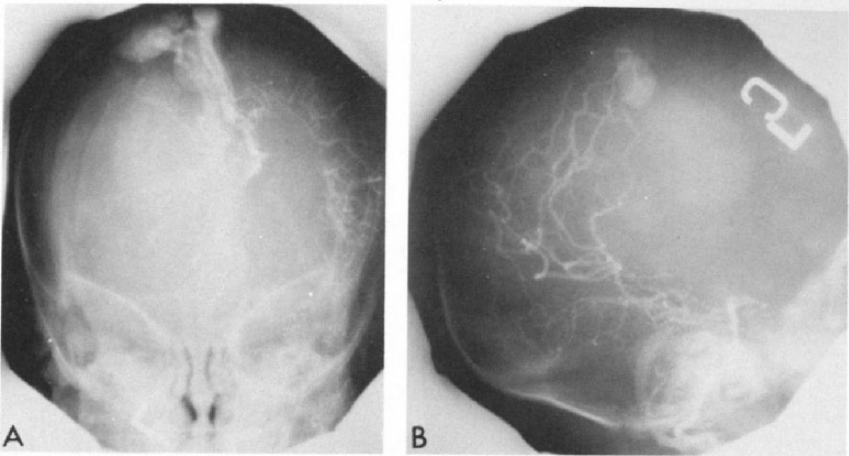


FIG. 16.7. (A) Left carotid and (B) vertebral angiograms of same patient as in Figures 16.5 and 16.6.

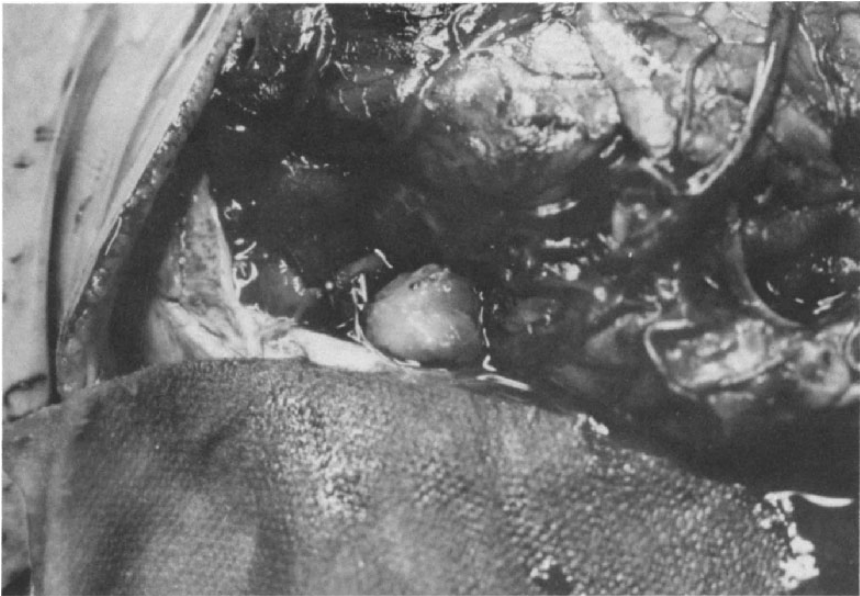


FIG. 16.8. Operative photograph showing superficial sac of AVM shown in Figures 16.6 and 16.7.

developed seizures. There was an angioma on the right parietal scalp (Fig. 16.5). Her right carotid arteriogram (Fig. 16.6) showed a trilobed sac. Two small superficial sacs were recipients of several arterial feeders from the carotid and also the vertebral arteries (Fig. 16.7). The larger sac

was apparently a blind pseudoaneurysm of one of the others. At craniotomy it was apparent that there had been extensive hemorrhage into the right hemisphere, presumably venous in origin, for damage was quite remote from the lesion (Fig. 16.8). It was eradicated without excision by securing the feeders at the smaller sac (Fig. 16.9). She was later shunted for a communicating hydrocephalus.

Some massive lesions are compatible with long life. A 14-year-old girl

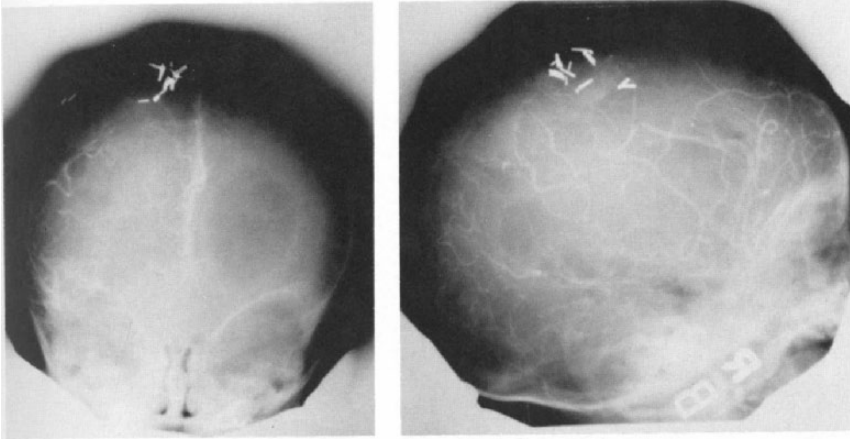


FIG. 16.9. Post-operative angiogram showing obliteration of the AVM. Note hydrocephalus.



FIG. 16.10. (A and B). Angiograms showing massive right hemisphere AVM in a 14-year-old girl. Venous drainage into the vein of Galen has enlarged it, but the shunt is diffuse and intracortical.



FIG. 16.11. CSF rhinorrhea in girl shown in Figure 16.10; hydrocephalus secondary to vein of Galen enlargement produced spontaneous, intermittent leak.

presented with headache and was found to have an enlarged sella turcica. It was said that she had “cerebral palsy” because of a very slowly progressive hemiparesis. A loud bruit was heard over the right posterior cranium. Arteriograms (Fig. 16.10) showed that a massive malformation occupying much of the right hemisphere secondarily involved the vein of Galen. Rhinorrhea (Fig. 16.11) prompted a shunting procedure. She is little changed 12 years after discovery of this inoperable lesion which has never bled. Her bruit has actually lessened. Although blood is shunted into the vein of Galen which is greatly enlarged, this is not an example of the operable primary malformation involving that vein.

Another girl (CD #83-52-03) suffered a subarachnoid hemorrhage at age 21. She was found to have an occipital AVM (Fig. 16.12). It was

resected using loupes and bipolar forceps without leaving any residual neurological deficit.

One AVM will always be remembered. A 16-year-old boy suffered an intracranial hemorrhage which led to arteriographic studies (Figs. 16.13 and 16.14) made shortly after his first hemorrhage. Plans had been made at another center for an elective stereotactic ablation of the lesion. Prior to that appointment, however, the boy had a lesser hemorrhage from which he recovered readily. The parents were understandably quite concerned and sought help elsewhere. It was elected first to perform a pneumoencephalogram and with air still in the ventricles to repeat the arteriographic studies to show the relationship of the lesion to the ventricle to aid in its localization. To our great surprise and joy, the

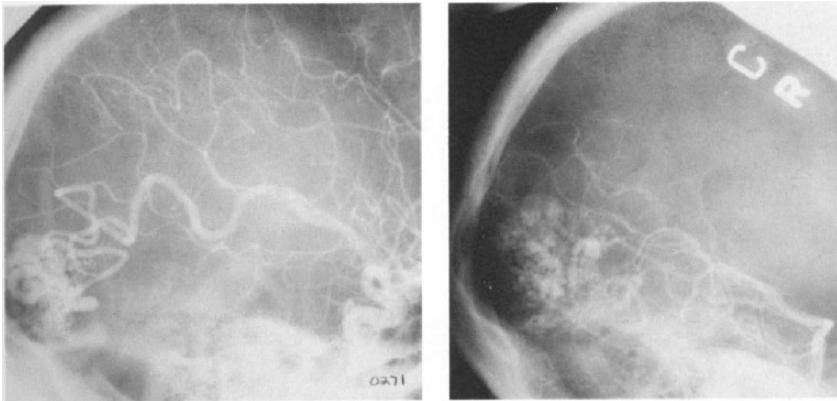


FIG. 16.12. Occipital AVM in a 21-year-old girl (C.D. #83-52-03), resected without neurological deficit. Blood supply was from carotid and vertebral circulation.

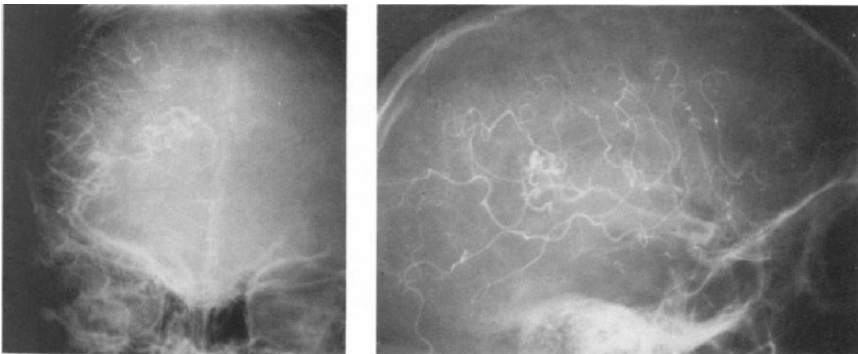


FIG. 16.13. Small deep AVM in dominant hemisphere of a 16-year-old boy. Hemorrhage prompted this first study. Arterial phase.

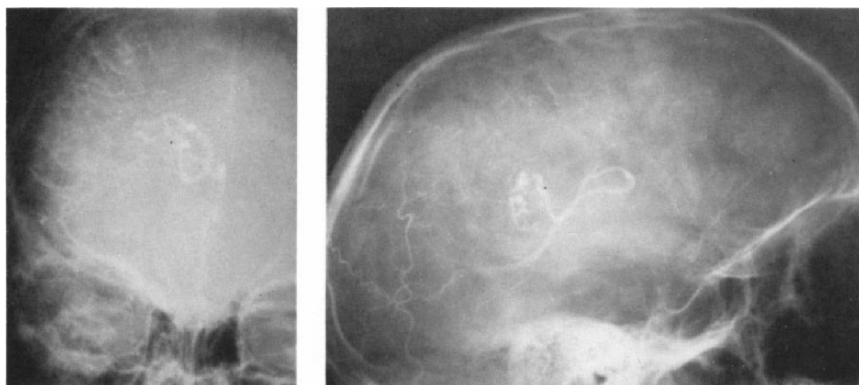


FIG. 16.14. Same patient as in Figure 16.13, venous phase.

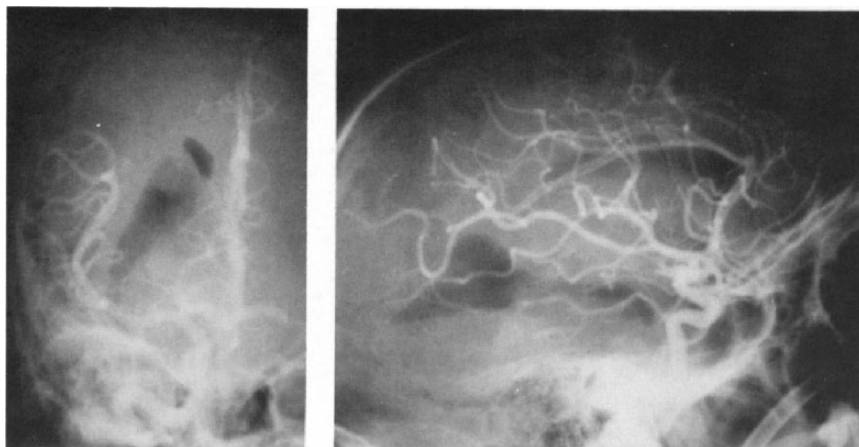


FIG. 16.15. Combination pneumoencephalogram and arteriogram in same patient as in Figures 16.13 and 16.14, after second hemorrhage had destroyed the malformation. Arterial phase.

malformation had disappeared (Figs. 16.15 and 16.16)! Neither we nor the parents could believe this good fortune so the arteriograms were repeated 6 months later. Still no sign of the malformation! The second bleeding episode apparently destroyed it completely.

In the cooperative study (17) of 52 children presenting with subarachnoid hemorrhage, 29% were found to have AVMs.

In the Children's Hospital Medical Center experience there have been 65 patients (Table 16.1). In his text *Neurosurgery of Infancy and Childhood* (19), Dr. Matson reported 34 confirmed AVMs between 1952 and 1966. These were exclusive of primary malformations of the vein of Galen, which were dealt with separately, as were post-traumatic

malformations, and the cryptic malformations which he considered hamartomas.

To this group has been added another consisting of 31 patients treated in the period 1967 to the present, October 1975, making a total series of 65 patients. The mortality rate associated with ligation or excision of the lesion has fallen from 24% to zero. The mortality rate in the group not dealt with surgically except by shunt has fallen from 53 to 7.5%. What this really means is hard to know without studying each patient's history and each patient's lesion.

Case 2. Shoveling snow; collapsed. This 11 2/12-year-old boy was shoveling snow when he collapsed with a headache and soon became stuporous. Arteriograms showed a deep

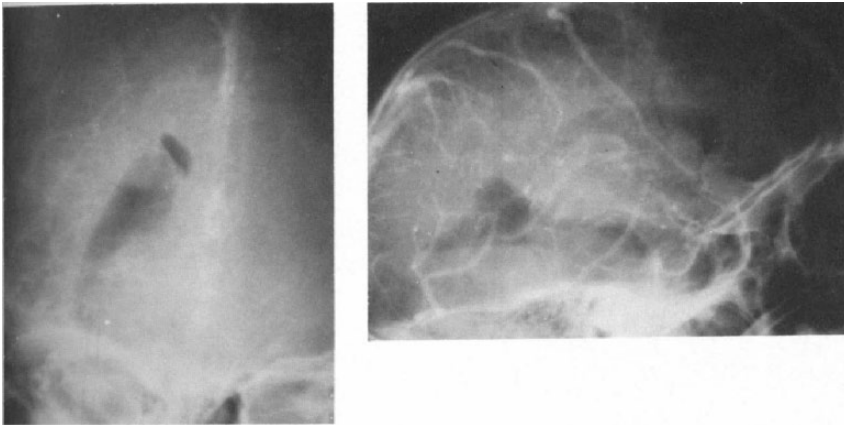


FIG. 16.16. Same patient as in Figures 16.13-16.15, venous phase. No sign of malformation.

TABLE 16.1.

Summary of Intracranial Arteriovenous Malformations Diagnosed at the Children's Hospital Medical Center, Boston, 1952-1975. Cryptic Microscopic and Primary Vein of Galen Malformations are not Included.

| PATIENTS | OPERATED | | | UNOPERATED | |
|-----------------|----------|---------------------------|---------|-----------------------------|------|
| | LIGATION | CLOT ONLY | FEEDERS | SHUNT | NONE |
| 34 1952-1966 | 12 | 4 | 1 | 3 | 14 |
| | | MORTALITY 24 % | | MORTALITY 53 % | |
| 31 1967-1975 | 15 | 0 | 1 | 1 | 14 |
| | | MORTALITY 0 | | MORTALITY 7.5 % | |
| 65 | 33 | TOTAL OPS, MORTALITY 12 % | | 32 PATIENTS, MORTALITY 34 % | |

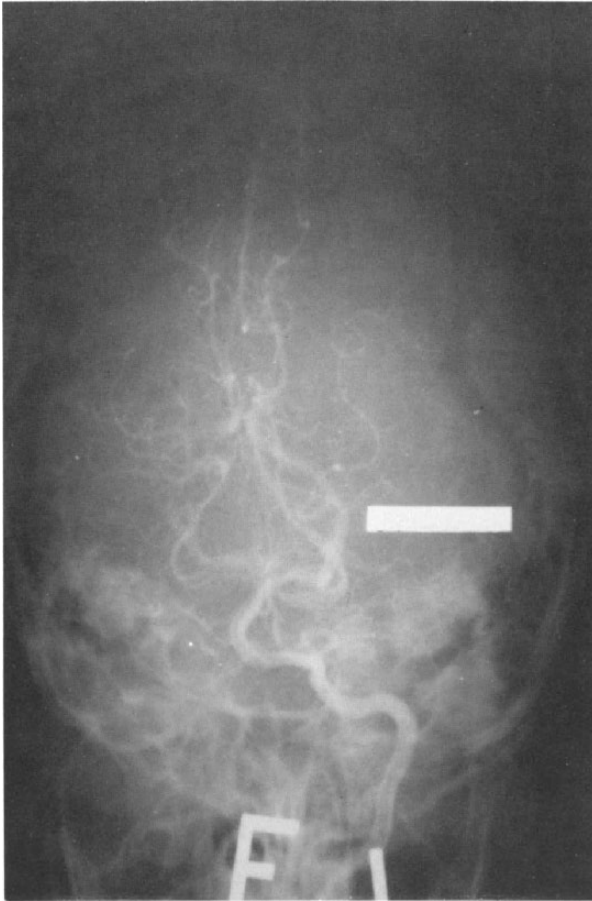


FIG. 16.17. Case 2. Arteriograms show a deep posterior parietal mass but no abnormal vasculature. Anteroposterior view.

posterior mass (Fig. 16.17) but no abnormal vasculature (Fig. 16.18). Clot was removed from the area within 24 hours of his first symptoms. He never awakened and died at the end of the week.

At autopsy, a microscopic AVM (Fig. 16.19) was found 1 cm. deeper than a piece of Gelfoam which had been placed on the wall of the cavity containing the clot.

Diagnosis: Cryptic vascular hamartoma of the parietal lobe.

All such lesions in our series have presented with hemorrhage, usually massive and sometimes fatal. The lesion has not been demonstrable angiographically. Usually the presence of a large amount of clot demands evacuation at which time the malformation may be found in the specimen. Repeated smaller bleeding episodes have been documented from such lesions and have been very puzzling. A 19-year-old girl had

three episodes over a period of as many years which were thought by their subtle and rapidly clearing nature to be indicative probably of multiple sclerosis (1). On the occasion of the third episode, however, unexpected xanthochromia of her spinal fluid prompted a pneumoencephalogram which showed a mass in her left thalamus (Fig. 16.20). Transparietal, transventricular exploration showed that this mass was only blood (Fig. 16.21). It was elected not to biopsy the wall of the cavity in this important area so what is left of the presumed malformation remains cryptic. Nine years later she is well, almost normal neurologically, is married, and has one child. Such malformations may be infratentorial also. One 13-year-old girl had numerous small bleeding episodes over a 2-year period, and many studies revealed no source. She ultimately died from a more massive hemorrhage. At autopsy a tiny subependymal malformation was found in the floor of the fourth ventricle (Fig. 16.22).

Case 3. *Minor dashboard bump; headache and hemiparesis.* A 2 1/12-year-old girl bent over to pick up an object which had fallen to the floor of her parents' Volkswagen. On the way up she bumped her head on the dashboard with no immediate sequela.

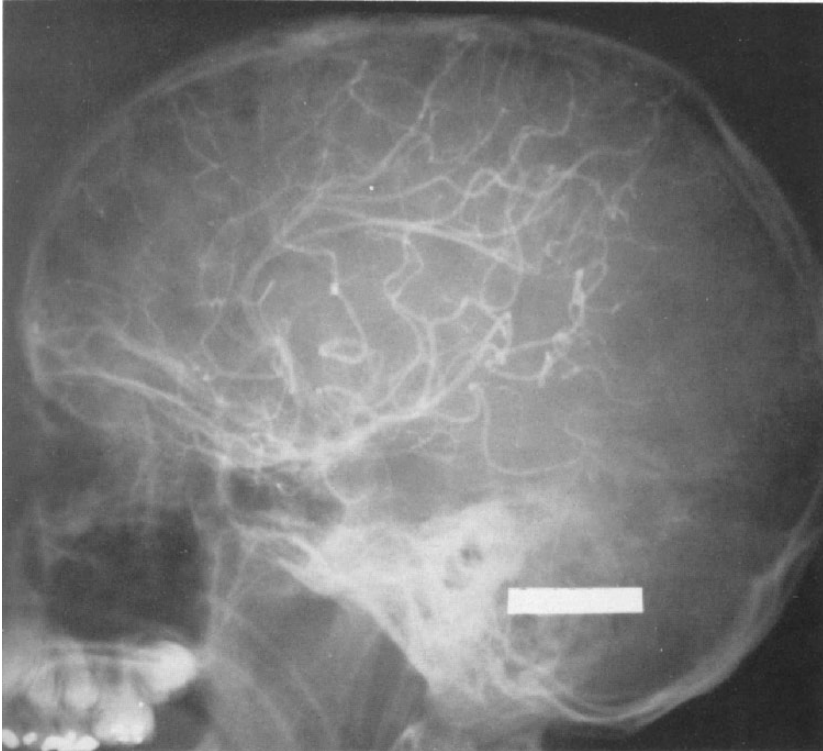


FIG. 16.18. Case 2. Lateral view.

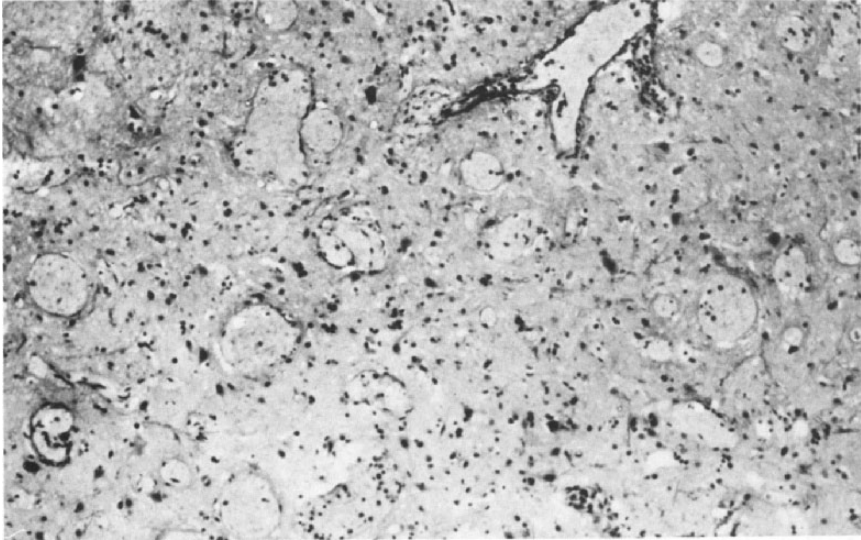


FIG. 16.19. Case 2. Microscopic arteriovenous malformation found at autopsy.

That night, 6 hours later, she awakened with severe headache, vomited, and developed a left hemiparesis. She had a history of generalized febrile convulsions. There was a "port wine" stain on the parietal scalp. She was still being treated with ampicillin because of a respiratory infection and otitis of 1 week's duration. A cousin had died in childhood because of a ruptured intracranial aneurysm.

A computerized axial tomography (CAT) scan showed right temporal lobe clot and an arteriogram showed an aneurysm of the middle cerebral artery deep to the clot (Fig. 16.23). She had a respiratory arrest which was fortunately reversible but which prompted emergency evacuation of the clot at which time the aneurysm was clipped. She survived. Four months later her hemiparesis was still improving.

Diagnosis: Congenital berry aneurysm of the middle cerebral artery.

In the cooperative study (17), of 52 children under 15 years presenting with subarachnoid hemorrhage, 27% were found to have aneurysms. The youngest child reported in whom an aneurysm has been successfully dealt with was 4 weeks old (13). The Children's Hospital Medical Center surgical experience with aneurysms over a 21-year period is shown in Table 16.2. The youngest patient was 16 months old; his middle cerebral aneurysm was clipped, but he did not survive. The youngest to survive rupture and surgery was 23 months old; the aneurysm was on the posterior inferior cerebellar artery.

The distribution of aneurysms, their gross appearance, and the relation to parent vessels is similar to those found in adults. Even an arteriosclerotic "sentinel plaque" was found in a 6½-year-old boy's carotid artery at the base of the bifurcation aneurysm.



FIG. 16.20. Pneumoencephalogram of a 19-year-old girl thought to have multiple sclerosis. Mass lesion occupies left thalamus.



FIG. 16.21. Operative photograph of girl shown in Figure 16.20. Thalamus contained old, unclotted blood.

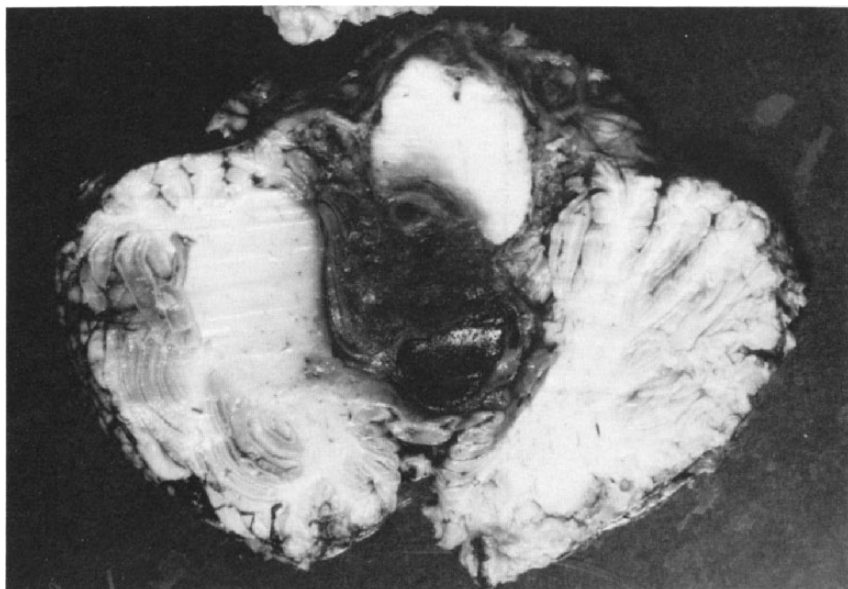


FIG. 16.22. Autopsy specimen of 13-year-old girl who died of hemorrhage from cryptic AVM in floor of 4th ventricle after many lesser hemorrhages.

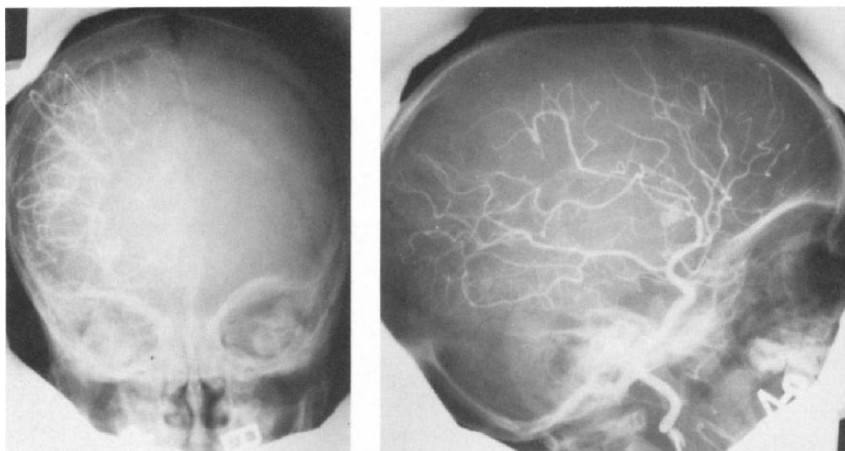


FIG. 16.23. Case 3. Right carotid arteriogram of 2½-year-old girl who hemorrhaged from middle cerebral aneurysm.

Spasm is just as prevalent in children as in adults. A 5½-year-old boy who bled from his apical basilar aneurysm (Fig. 16.24), showed marked widespread spasm which produced devastating neurological deficits (Fig. 16.25). He had a second aneurysm on his left posterior cerebral artery.

After waiting several weeks for the spasm to subside, the aneurysms were coated. He died several years later of complications of shunted hydrocephalus.

In children, other undiscovered congenital anomalies may be present, such as coarctation of the aorta and polycystic kidneys, and a search

TABLE 16.2.

Summary of Congenital Intracranial Aneurysms Diagnosed at the Children's Hospital Medical Center, Boston, 1954-1975

| | | |
|---------------------------|-------|----------|
| INTERNAL CAROTID | ----- | 5 |
| ANT. COMM., ANT. CEREBRAL | ----- | 8 |
| MIDDLE CEREBRAL | ----- | 4 |
| VERTEBROBASILAR | ----- | <u>3</u> |
| TOTAL | | 20 |

AGES 1 4/12 - 17 7/12 YRS.

19 OPERATED UPON

16 SURVIVORS

16 % OPERATIVE MORTALITY

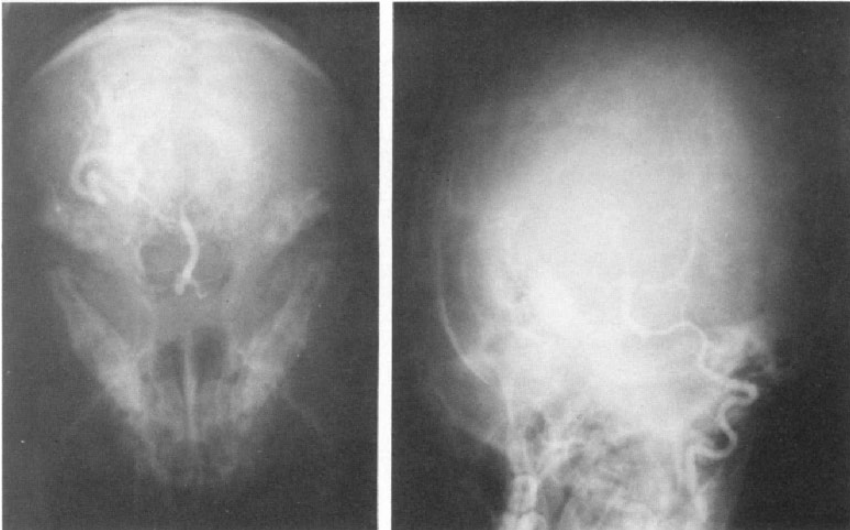


FIG. 16.24. Two aneurysms on the posterior circulation of a 5½-year-old boy. Apical basilar aneurysm had bled; second on left posterior cerebral had not. Oblique and basal views.

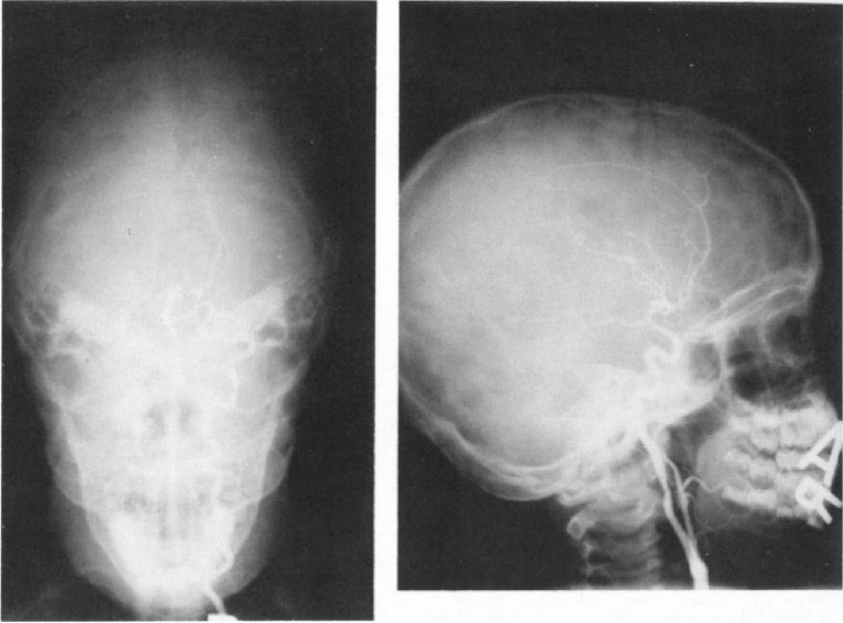


FIG. 16.25. Same boy as in Figure 16.24, showing marked spasm. Anteroposterior vertebral and lateral right carotid injections.

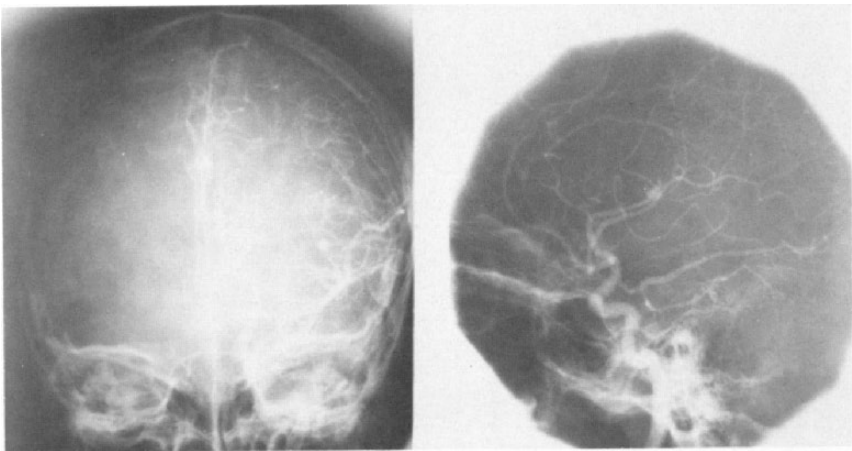


FIG. 16.26. Case 4. Left carotid arteriogram of 9½-year-old girl, showing mycotic aneurysm on middle cerebral artery.

should be made for these conditions whenever a child presents with an intracranial aneurysm.

In newborn infants, massive intracranial hemorrhage may occur from the choroid plexus or, in premature infants, from the ganglionic

eminence, an area on the ventricular surface of the caudate nucleus. Both regions have been shown by Gilles (9) to be high in fibrinolytic activity which may propagate hemorrhage once it has occurred. The great susceptibility of premature and newborn infants to hemorrhage of this type is not fully understood. High venous pressure and venous stasis resulting from immaturity of the pulmonary vascular bed or from hyaline membrane disease may play a role. The neurosurgeon must be alert to rule out subdural hematoma caused by head molding at delivery and also retrocerebellar hemorrhage from tears of the venous sinuses (10). If the infant survives the bleeding episode, hydrocephalus or chronic subdural effusions may require action.

Children may suffer from spontaneous intracranial hemorrhage because of congenital abnormalities of the clotting mechanism, such as in hemophilia (15, 20, 23) or disease processes such as leukemia (21). Idiopathic thrombocytopenic purpura has been reported in children (18). Sickle cell disease can lead to intracranial hemorrhage as well as occlusion of intracranial vessels (2, 6).

Case 4. *Meningitis, endocarditis, osteomyelitis, and sudden headache.* A 9½-year-old girl developed staphylococcus meningitis. During therapy she was found to have acute bacterial endocarditis and osteomyelitis of one metacarpal bone. This area was drained surgically. One month after the onset of her initial symptoms, she had a spontaneous intracerebral hemorrhage. Arteriography (Fig. 16.26) that day revealed an aneurysm of the left middle cerebral artery with proximal spasm. She was treated with epsilon-aminocaproic acid (Amicar, Lederle, Pearl River, N. Y.), 1.5 gm. intravenously every 2 hours, a dose of 0.7 mg. per kilogram per day.



FIG. 16.27. Case 5. Cutaneous hemangioma at base of neck.

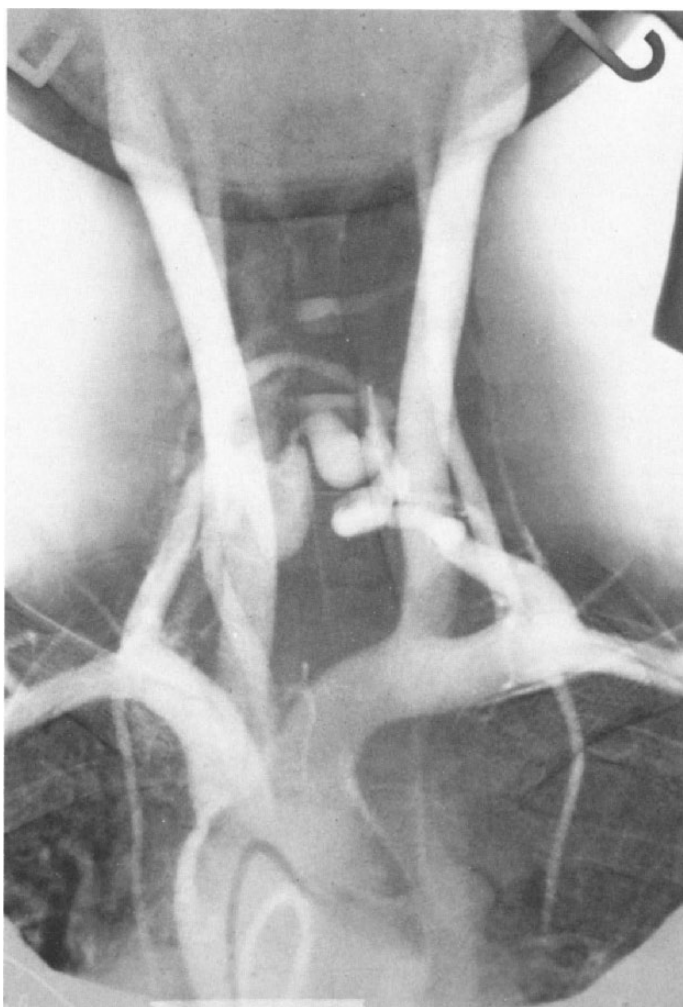


FIG. 16.28. Case 5. Arch aortogram. Intraspinal AVM is fed by a cervical artery on each side.

She improved neurologically and her CSF and blood cultures became negative. A second arteriogram suggested enlargement of the aneurysm and therefore craniotomy was performed 4 weeks after the hemorrhage. The pia arachnoid in the Sylvian fissure was significantly thickened and densely adherent to the middle cerebral vessels. An aneurysm 5 mm. in diameter was found, dissected free, and coated with plastic by the technique of Selverstone (24, 25). The girl has done well and 2 years later is normal except for a seizure tendency and some learning disability.

Diagnosis: Mycotic aneurysm, middle cerebral artery.

Case 5. *Riding bike, severe headache, tingling arms, weak legs.* This 8 10/12-year-old boy was riding his bike when he developed a severe headache which promptly spread to his

neck and back. He noticed a heaviness of his arms and could no longer easily ride. He walked the bike home. The headache persisted until he arrived in the emergency ward where he was found to have nuchal rigidity but was neurologically normal except for an equivocal response in the left great toe. Lumbar puncture yielded grossly pink fluid, a pressure of 210 mm., 39,000 red cells per cubic millimeter and a protein of 109.5 mg. per 100 ml. There were multiple pink flat hemangiomas of his trunk and extremities, the largest of which was at the base of his neck posteriorly (Fig. 16.27). He gave a history of having been hit on the head by a friend 2 days prior to admission.

By the next day his headache was gone. Skull films, EEG, technetium scan, and flow studies were all normal. On the following day, cerebral angiography was performed and was completely normal. Because of the cervical hemangioma, an arch study was done and the neck filmed (Fig. 16.28). An intraspinal AVM was demonstrated. It was fed by two enlarged cervical arteries (Fig. 16.29) arising from the region of the thyrocervical trunk on each side just distal to the origins of the vertebral arteries. A loud bruit was then noted over the C-7 area!

An elective procedure was carried out 2 weeks later. It was very much a joint effort between the patient's neurologist, the neuroradiologist, and the neurosurgeon. With the boy sedated but awake, the procedure began in the x-ray department. Through the right axillary artery a #6 French catheter with a balloon near its tip was passed into the right feeding artery. Through the left femoral artery, a similar catheter was passed into the left feeding vessel. A third catheter was passed through the right femoral artery to the aortic arch.

While the boy's neurological state was carefully and frequently tested, first one and then the other balloon was inflated with Renografin. The boy did not show any change, except that his bruit disappeared. Test injections into the arch showed no filling of the AVM (Fig. 16.30). The boy was observed for a total of 1 hour with the balloons inflated without showing any neurological change.

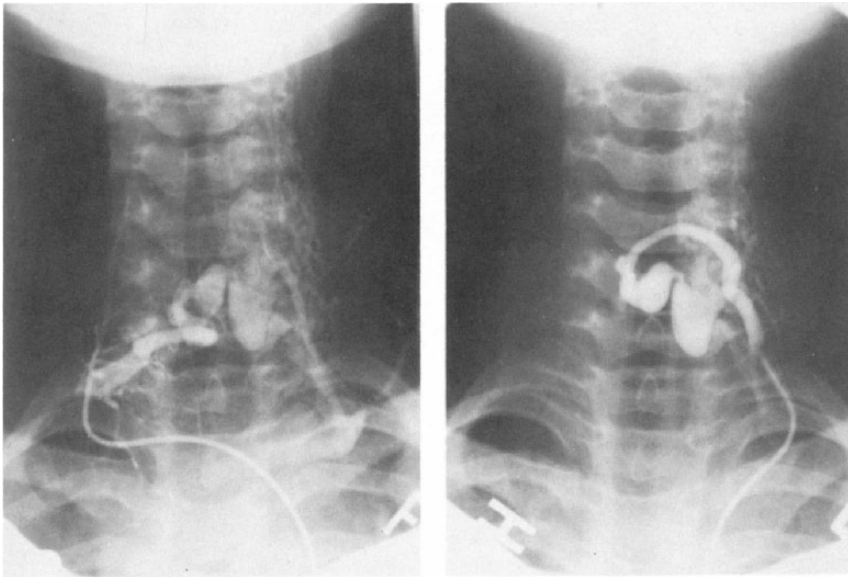


FIG. 16.29. Case 5. Detail of left and right feeding vessels.

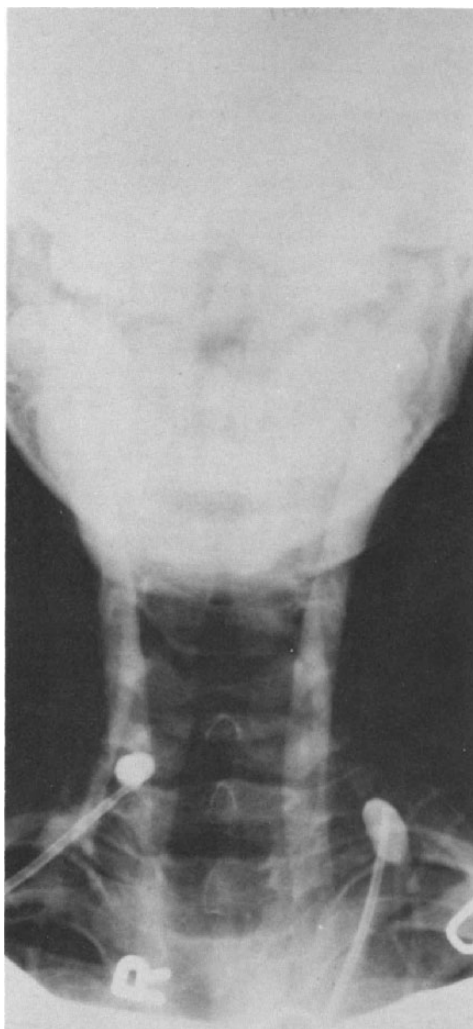


FIG. 16.30. Case 5. Arch aortogram after occluding both feeding vessels with balloon catheters. AVM no longer fills.

Next, in the operating room, catheters still in position and balloons inflated, the boy was anesthetized and each feeding vessel isolated and ligated. The balloon catheters were felt to be invaluable in the localization of the feeding vessels. The bruit has not recurred and the boy has remained well and fully active.

Follow up angiography was performed about 5 months following surgery. Injection of dye near the origin of the right thyrocervical trunk showed no evidence of the malformation. To our dismay, a similar injection on the left side showed the feeding vessel intact and the malformation still filling from it! Whether the balloon catheter changed its position during the trip to the operating room or whether the surgeon imagined he felt the catheter in the

vessel he ligated will always be a mystery. At this time, our plans are to embolize the shunt through the remaining feeding vessel.

Diagnosis: Intraspinal AVM.

Case 6. *Episodic vomiting, recent papilledema, acute hemiparesis.* An 11 9/12-year-old

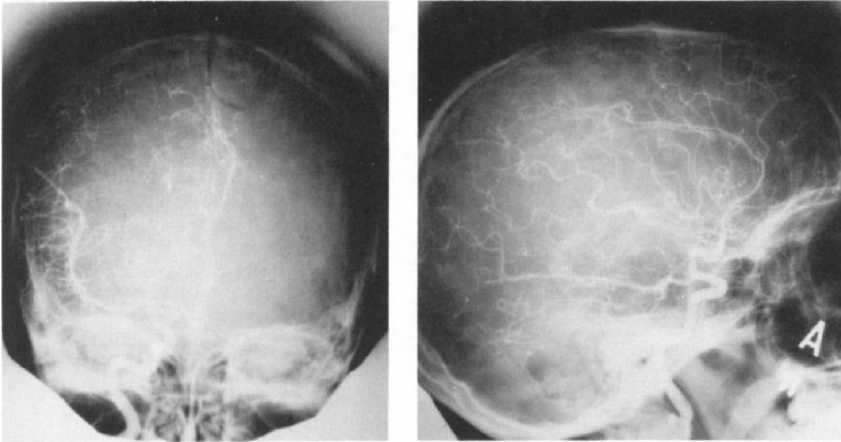


FIG. 16.31. Case 6. Right carotid arteriogram showing mass effect but no abnormal vasculature.

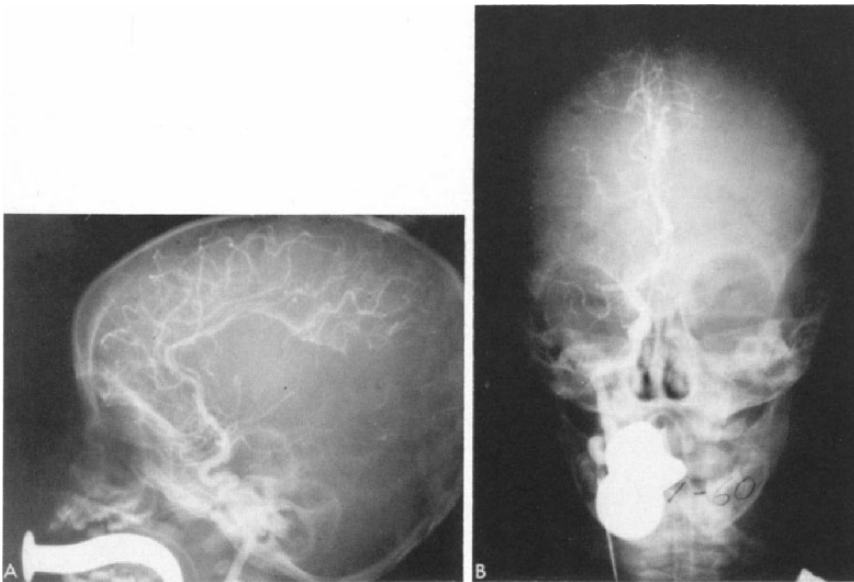


FIG. 16.32. Case 8. Lateral (A) and anteroposterior (B) views of right carotid arteriogram, showing occlusion of the middle cerebral artery just distal to a small temporal branch (From J. Shillito: Carotid arteritis; a cause of hemiplegia in childhood. *J. Neurosurg.*, 21: 540-551, 1964.)



FIG. 16.33. Case 8. Operative photograph showing occluded middle cerebral artery. Right subfrontal approach (From J. Shillito: Carotid arteritis; a cause of hemiplegia in childhood. *J. Neurosurg.*, 21: 540-551, 1964.)

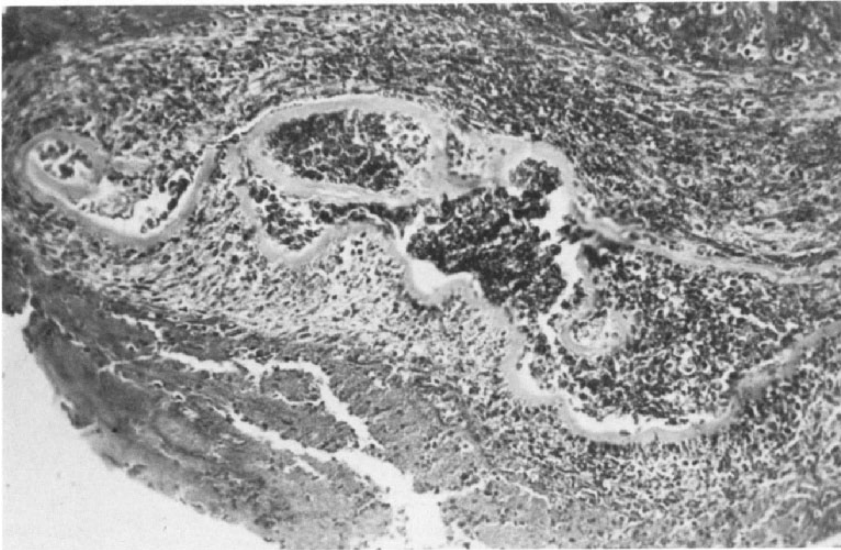


FIG. 16.34. Case 8. Microscopic view of specimen removed. Thrombus is surrounded by what appears to be displaced internal elastic membrane and a surrounding zone of arteritis (From J. Shillito: Carotid arteritis; a cause of hemiplegia in childhood. *J. Neurosurg.*, 21: 540-551, 1964.)

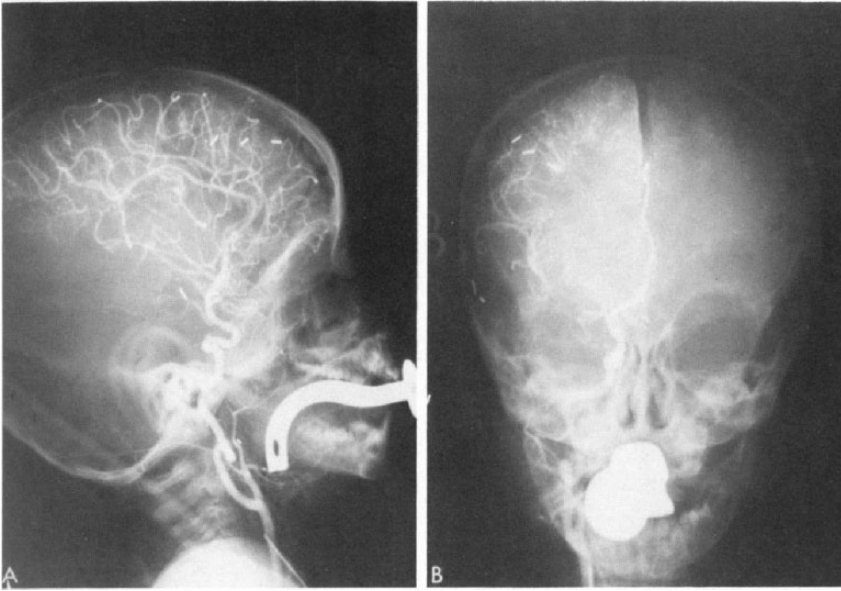


FIG. 16.35. (A and B). Case 8. Postoperative arteriograms show persistence of the block at site of arteriotomy, but good collateral filling of middle cerebral artery (From J. Shillito: Carotid arteritis; a cause of hemiplegia in childhood. *J. Neurosurg.*, 21: 540-551, 1964.)

CONTRIBUTING FACTORS, CEREBRAL ARTERIAL DISEASE
25 CHILDREN

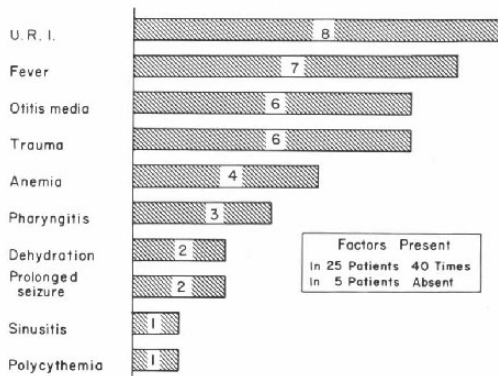


FIG. 16.36. Possible contributing factors in cerebral arterial disease of 25 children (From J. Shillito: Carotid arteritis; a cause of hemiplegia in childhood. *J. Neurosurg.*, 21: 540-551, 1964.)

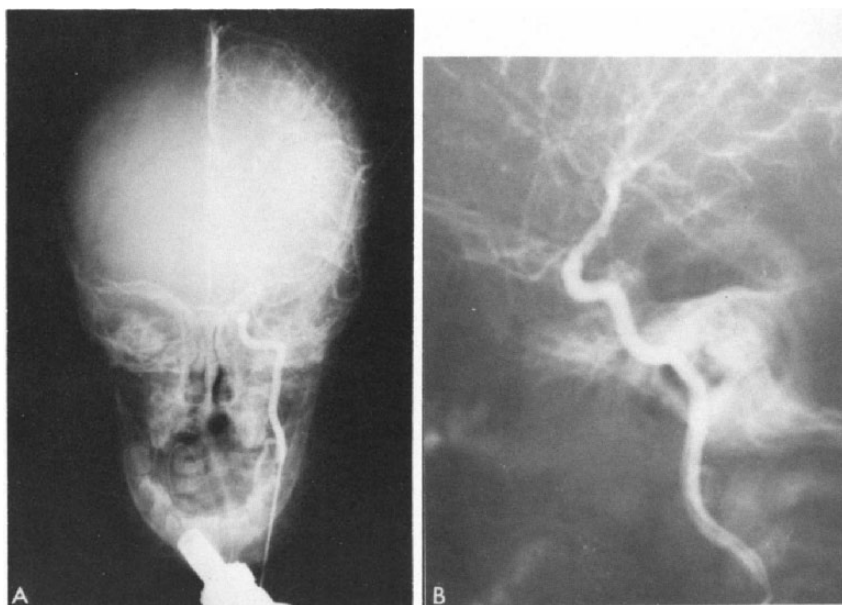


FIG. 16.37. (A and B). Left carotid arteriogram of a 1 2/12-year-old girl who had a right hemiparesis. Flow is retarded at carotid intracranially, where vessel appears irregular and somewhat narrowed (From J. Shillito: Carotid arteritis; a cause of hemiplegia in childhood. *J. Neurosurg.*, 21: 540-551, 1964.)

boy developed a right hemiparesis within the 1 1/2 hours necessary to get from his pediatrician to the neurosurgeon. The referral was made because of a change in the appearance of his optic discs compared to examination 1 week earlier and because of some difficulty which he was experiencing with vision. He had had several episodes of vomiting during the previous 2 months. On the first occasion, gastroenteritis was prevalent and he was thought to have this. Ten days prior to admission the second transient episode prompted a gastrointestinal series which was normal. His third episode prompted the current visit to the pediatrician.

When first seen by the neurosurgeon he had 4 diopters of papilledema, no hemorrhages, and enlarged blind spots. New in the past 1 1/2 hours were the neurological findings: a left hemiparesis, left paresthesias with extinction on sensory examination, and an upgoing left toe. He could not stand and moved his left arm only by lifting it with his right. Dexamethasone (Decadron, Merck, Sharp & Dohme, West Point, Pa.) and mannitol were started immediately and a technetium scan and arteriogram performed. These showed an avascular mass deep in the right parietal area (Fig. 16.31). Because of worsening of his condition a few days after admission, an emergency craniotomy was performed which revealed a large glioblastoma, partly cystic and partly within the right lateral ventricle. Strangely enough, there was no sign of recent hemorrhage nor was that ventricle enlarged as though acutely obstructed. Subtotal resection of the tumor was followed by radiation therapy. The boy died 9 months later.

Diagnosis: Malignant tumor of the right cerebral hemisphere.

Brain tumors have mimicked strokes by bleeding or by acutely obstructing a lateral ventricle. This tumor appeared to have done

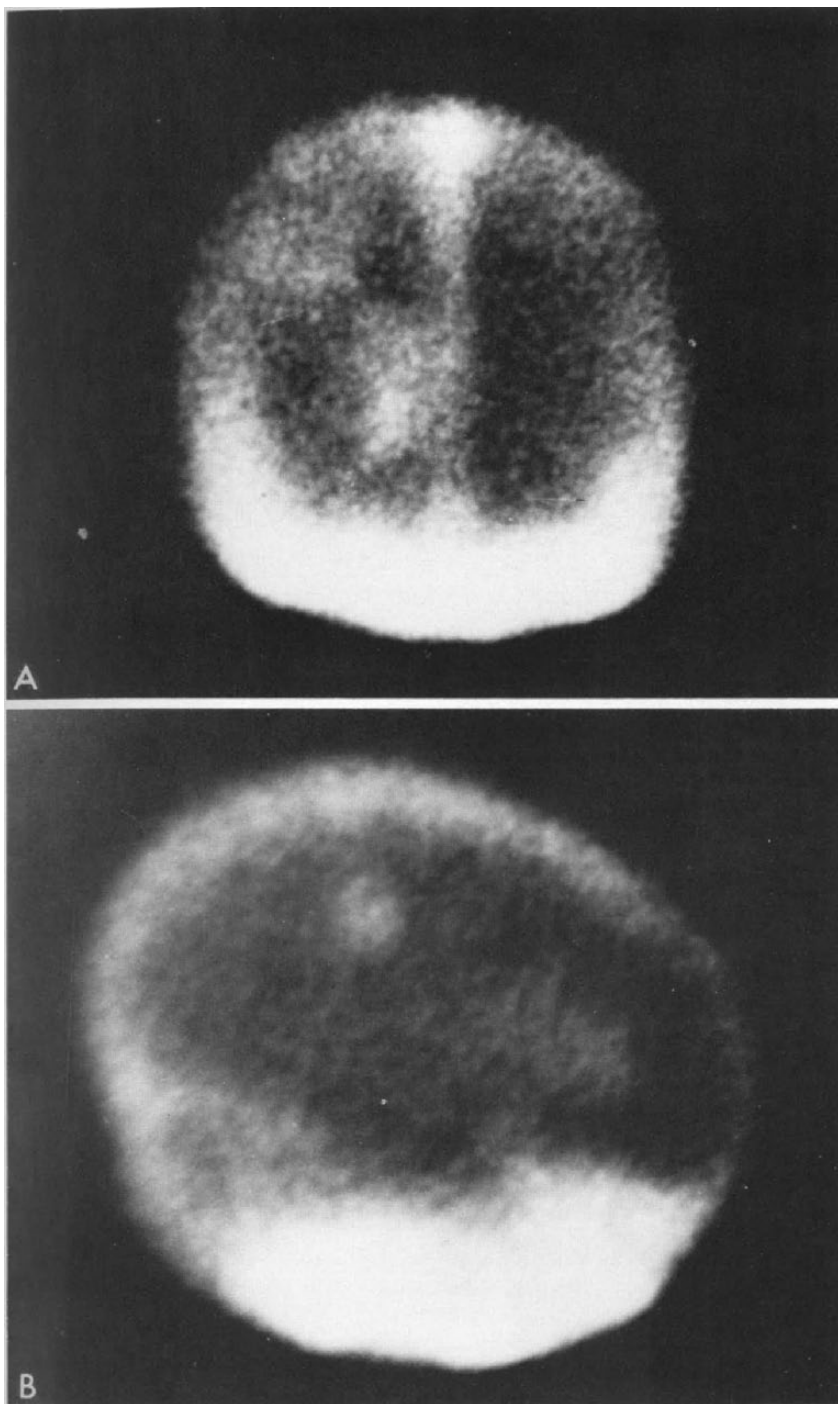


FIG. 16.38. (A and B). Case 9. First technetium brain scan, showing two lesions on the right side.

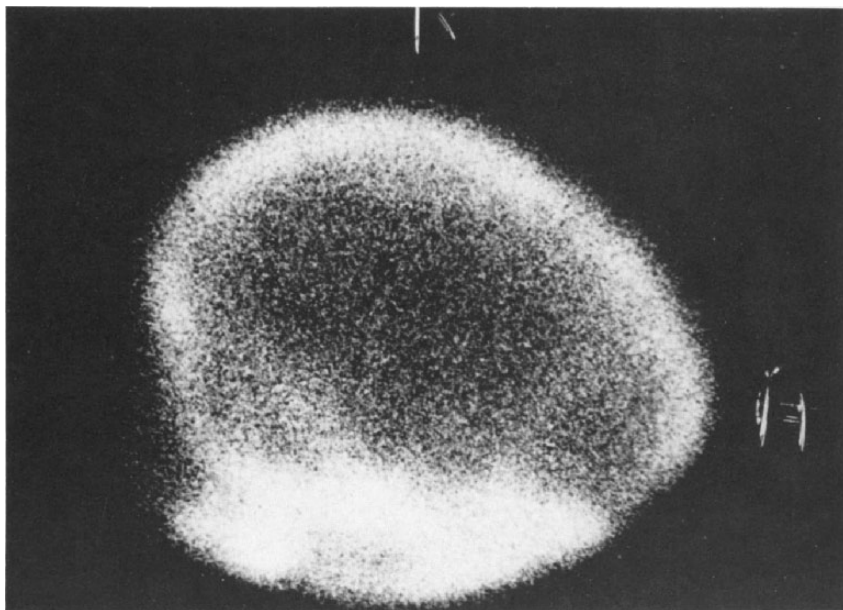


FIG. 16.39. Case 9. Second Tc scan 3 days later—previous lesions are not seen!

neither. Usually the tumors which bleed are intraventricular, such as the choroid plexus papilloma, malignant, or both. Recently, acute cerebellar hemorrhage in a child was found to have originated from a malignant tumor. These possibilities must be kept in mind when planning studies for the elucidation of what may appear superficially to have been a cerebrovascular accident.

Case 7. *Catching football; fell hemiplegic.* An 11 2/12-year-old boy was running to catch a football thrown by his father when he suddenly fell to the ground. He had not tripped and had no headache but said his left leg had simply buckled and given way. He tried to stand but his leg would not support him. He could not make a fist with his left hand. There had been no alteration in consciousness and no seizure activity had been witnessed.

Three hours later in the hospital he could just wiggle his left big toe and could get his left arm up over his head briefly. Throughout the night his hemiparesis continued to improve. He admitted to putting his head down on his desk at school the day before because he just didn't feel quite right and to having had a few ill defined headaches during the past few months.

The next day he was further improved. A technetium static scan and flow study both were normal. A cerebral angiogram was completely within normal limits, including the vessels in the neck! The boy was discharged and within 10 days he was back to normal.

Two months later he experienced dizziness, tingling of his *right* arm and leg, and weakness of the *right* side. The left side, which had been previously involved, remained completely normal. He could walk but limped. By the time he arrived in the hospital, he was already improving. His lumbar puncture showed 57 red cells, 35 mg. per 100 ml.

protein, 1.2 mg. per 100 ml. gamma globulin. There was no evidence of cardiac vegetations or intraventricular myxoma by echocardiogram. Technetium scan and flow studies again were normal. A CAT scan was normal. The episode cleared slowly with a little residual even 2 months later. He has presented once since that time with headache but no new neurological deficit.

Diagnosis: Hemiplegic migraine.

This has been reported in children under 3 years of age (32).

Case 8. Awoke with weak arm. A 1-year-old boy awoke on the day prior to admission with a weak left arm. During the previous 2 weeks he had had a minor fall without head injury and a nonproductive cough. On admission he had a left hemiparesis, slight reddening of the right ear drum, and a hemoglobin of 7.4 gm. per 100 ml.

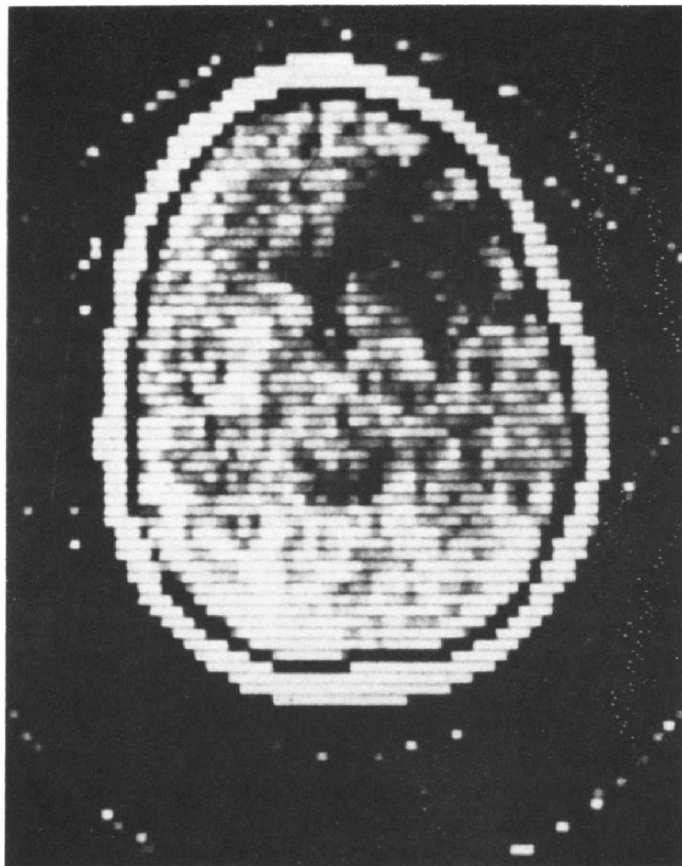


FIG. 16.40. Case 9. First CAT scan, at about the time of negative Tc scan, shows a heterogeneous mass of low mv in the right frontal lobe, displacing the ventricular system to the left and partially amputating the right frontal horn.

After transfusion, arteriography was performed on the 2nd hospital day. The right middle cerebral artery was occluded just distal to a small temporal branch (Fig. 16.32). At craniotomy immediately thereafter, a firm white obstruction was found straddling a major bifurcation of the middle cerebral artery (Fig. 16.33). It was removed with some difficulty; the arterial wall was thickened and the mass was adherent to it. After closing the arteriotomy, blood filled the distal branches which then pulsated. Microscopically (Fig. 16.34), it appeared that part of the wall of the vessel had been removed in the specimen, for one area showed thrombus in a narrowed lumen where an unusual density of round cells and some neutrophils surrounded a stripped internal elastic membrane. This was considered either reaction to a clot present for several days or a primary process which caused the thrombosis. It is quite similar to reported cases of cerebral-dissecting aneurysms. Arteriography, 3 weeks postoperatively (Fig. 16.35A), demonstrated excellent filling of all branches of the middle cerebral artery, but closer inspection of the films, particularly the anteroposterior view (Fig. 16.35B), showed that the obstruction had recurred at the operative site and demonstrated a remarkable collateral circulation *via* the small temporal branch of the middle cerebral artery and *via* the anterior cerebral circulation. The child has recovered with minimal dysfunction of the left hand.

Diagnosis: Arteritis, etiology unknown.

In 1964, 25 patients from the Children's Hospital Medical Center files were reported who had had sudden ischemic strokes followed by arteriographic study (26). Twenty showed total arterial occlusion, 4 partial occlusion, and the study in one case suggested recannulization. In all but 5 of these patients there was some factor or combination of factors that might have contributed to inflammation of the carotid artery near the base of the skull (Fig. 16.36). These included upper respiratory infection, fever, otitis media, trauma, pharyngitis, and sinusitis. Arteritis was suggested by the radiographic appearance of one carotid artery incompletely occluded intracranially just at the carotid syphon (Fig. 16.37), by surgical specimens, and by one autopsy specimen. Surgery in no case permanently relieved the obstruction but current microsurgical techniques were not then employed. Those few cases which have come to attention since that report have been diagnosed several days after the onset of symptoms and exploration has not been carried out.

There are other causes of vascular occlusion in childhood. Taveras (29) reported 10 cases of multiple progressive intracranial arterial occlusions, 5 of which were in children under 16 years of age, one as young as 7 months.

The strange "moyamoya" disease (28) with its blush of tiny vessels near the base of the brain resembling a puff of cigarette smoke may be, in fact, rich anastomotic pathways developing early in life in response to carotid stenosis or occlusion. Polyarteritis (8), sickle cell disease (27), systemic lupus erythematosus (12) all can produce symptoms of stroke. In teenagers, one must also consider vascular problems associated with drug abuse (16) and contraceptive medications (4).

Occlusion of cerebral veins and venous sinuses can occur in children for

many reasons (5, 14,): infection, anemia, and, particularly, congenital heart disease (31). An important warning: in children with cyanotic congenital heart disease, any sudden focal neurological deficit whether static or progressive must be considered as possibly due to a cerebral abscess (7) and studies planned to establish or rule out this diagnosis promptly and with safety (30).

We have found arteriography in the pediatric age group to be safe. Only in the presence of homocystinuria is the risk of cerebral angiography considered forbidding. There have been cases reported in which neurological worsening was apparently caused by cerebral angiography in children with multiple intracranial arterial occlusions and homocystinuria. We take the precaution of a urinary nitroprusside test before performing angiography on any child who is thought to have had

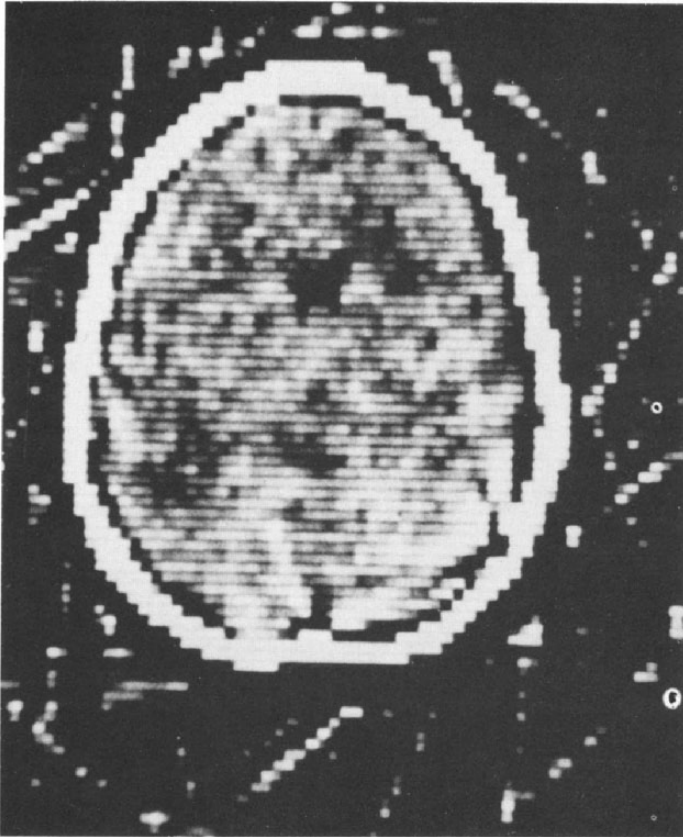


FIG. 16.41. Case 9. Second CAT scan 2 months later. Almost complete clearing of previous low density area. Ventricles have returned to their normal position.

an ischemic cerebrovascular accident. Angiography would not be performed in any child with the clinical features of homocystinuria (22).

Case 9. Recurrent occipital headaches, facial weakness. A 5 5/12-year-old girl had complained of occipital headaches several times during the preceding 5 months. Once, a month earlier, she rose from her nap drooling from the left corner of her mouth, seemed to be disoriented, vomited, and went back to bed again. After awakening 2 hours later, she appeared normal except for a fever which went as high as 102°F. and lasted for about 2 days. Two weeks prior to admission, she had again a severe occipital headache and a left facial weakness. She underwent several tests at that time, including an EEG and skull films, all of which were normal. Her symptoms subsided.

On the day of admission she again appeared with a left facial weakness, some awkwardness of left-sided coordination and was felt by some observers to have early papilledema. All tests including arteriograms were normal except a technetium brain scan which showed two areas of increased concentration of isotope in the right cerebral hemisphere (Fig. 16.38). The girl was placed on dexamethasone (Decadron), and a search was begun for a possible primary site of metastatic tumors. The search was not productive. A second technetium scan 3 days later was completely normal (Fig. 16.39)! A CAT scan, however, suggested a lesion deep in the right hemisphere (Fig. 16.40). The lesion was described as a heterogeneous mass of low μ in the right frontal lobe which displaced the ventricular system to the left and partially amputated the right frontal horn. It was felt to be consistent with a glioma. To define this in relation to the ventricular system, a pneumoencephalogram was performed. This showed only diffuse expansion of the deep portion of the right hemisphere. The spinal fluid protein was 20 mg. per 100 ml. and gamma globulin was normal. The girl was improving on dexamethasone (Decadron) and, because of this and the improving technetium scan, it was decided simply to observe her further. Two months later the CAT scan was much improved (Fig. 16.41), and the patient was normally active!

Four months after the first hospitalization, she had recurrence of her left occipital headache lasting 1 to 2 hours, this time with a *right* facial weakness and slurred speech. The function of her entire right side appeared to be somewhat abnormal and she was felt to have early papilledema. She was readmitted for study. A lumbar puncture was 220 mm. of water with a total protein of 25 mg. per 100 ml. and the gamma globulin again was normal.

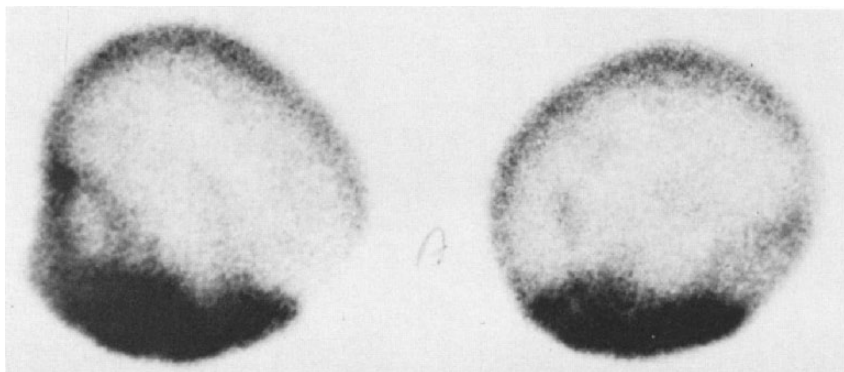


FIG. 16.42. Case 9. Third Tc scan at time of right-sided signs. Four lesions are suggested on the left side.

Technetium scan at this time showed *four* lesions in the left hemisphere (Fig. 16.42). She was again placed on dexamethasone (Decadron) and her neurological status and scans returned to normal. Two months later she complained of poor vision. There was no other neurological deficit. A central scotoma was found in her left eye and a lesser one in the right. A diagnosis of optic neuritis was made and her entire course now appears compatible with. . . .

Diagnosis: Multiple sclerosis.

SUMMARY

Nine cases have been presented in detail to illustrate some of the varied causes of sudden neurological deficit in childhood: arteriovenous malformation, cryptic hamartoma, berry aneurysm, mycotic aneurysm, intraspinal arteriovenous malformation, brain tumor, migraine, arteritis, and multiple sclerosis. The Boston Children's Hospital experience with aneurysms and intracranial arteriovenous malformation has been summarized.

It is noteworthy that a cutaneous hemangioma overlay one cranial and one intraspinal arteriovenous malformation.

One small but deep cerebral arteriovenous malformation apparently destroyed itself after its second hemorrhage.

Not only have multiple sclerosis and a brain tumor mimicked a vascular lesion, but a series of vascular accidents was misdiagnosed first as multiple sclerosis then as a thalamic tumor.

The many possible causes of childhood strokes has been thoroughly cataloged in the Report of the Joint Committee for Stroke Facilities in 1973 (11). Children may be more susceptible to strokes because of congenital abnormalities such as congenital heart disease, hemophilia, and sickle cell anemia, or by diseases which more commonly occur in this age group, such as leukemia.

The likelihood of brain abscess in cyanotic congenital heart disease is stressed. Arteriographic studies in our series have been safe; however, there have been reports of probable worsening of symptoms in children with multiple cerebral occlusive lesions in the presence of homocystinuria.

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