ARteriovenous ANeurysm of the Posterior Fossa in an Infant

REPORT OF A CASE

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Vascular malformations of the posterior fossa in infants are extremely rare. Dandy3 reported an infant of 1 month with an angiomata of the posterior fossa with extracranial and intracranial extension and development of hydrocephalus who subsequently died on attempted removal of the tumor. Richardson and Bagnall8 described a case of angiomata of the posterior fossa in a male 23 years of age whose symptoms were first noted at the age of 7 years. The patient died at the age of 23 years, and at autopsy an angiomatous malformation of the posterior fossa was found. Grotts4 reported a 2-year-old child, who died from a spontaneous hemorrhage, with a cavernous hemangioma of the pons found at autopsy. Olivecrona and Riives6 reported 60 cases of arteriovenous aneurysms out of 5206 verified tumors of the brain. Of these 60 aneurysms, only 2 were in the posterior fossa, and these 2 were in males aged 39 and 50 years. Clement et al.3 reported 2 cases of arteriovenous aneurysms in infants aged 5 weeks and 8 months. The first case was that of an infant who died at the age of 5 weeks; autopsy revealed an avoid aneurysmal dilatation at the junction of the free edge of the falx cerebri with the tentorium cerebelli. The second child had dilatation of the right sinus rectus, torcular Herophili, and lateral sinus as well as an arteriovenous aneurysm of the ampulla of Galen. These were proved by angiography. The findings were verified at autopsy.

The following case report is of a 2-month-old white male infant who had a spontaneous subarachnoid hemorrhage from an arteriovenous aneurysm of the posterior fossa which was diagnosed by vertebral angiography and successfully removed.

CASE REPORT

JG #174594. A 2-month-old white male infant was admitted to the Baptist Memorial Hospital on Nov. 18, 1954. He was referred to this hospital by Dr. J. N. Robinson, Columbus, Miss., and Dr. J. R. Mullens, Jr., West Point, Miss. The infant was a full-term fetus delivered under general anesthesia with no complications. He was apparently normal until 3 weeks before admission when he was said to have had convulsion with loss of consciousness. He was taken to Dr. Mullens, who referred the child to Dr. Robinson. A spinal puncture yielded xanthochromic fluid. The examination prior to this time had shown that the anterior fontanelle was tense and bulging. A pneumoencephalogram was performed by Dr. Robinson which showed slight ventricular enlargement and absence of air over the cerebral hemispheres.

Examination. The head was obviously enlarged. The occipitofrontal circumference of the skull measured 41 cm.; the anterior fontanelle was large and bulging. Lumbar puncture revealed slightly xanthochromic spinal fluid with 164 mg. per cent of total protein. Ventricular fluid was colorless.

Ventriculography revealed the lateral and 3rd ventricles to be symmetrically enlarged.

with no filling of the 4th ventricle or the aqueduct of Sylvius. The enlargement of the ventricles was noted to be greater than on the previous encephalogram. Two days following this pneumoencephalography was performed with 50 cc. of air to demonstrate the patency of the 4th ventricle and aqueduct. The lateral ventricles were well filled, as was the 3rd ventricle. Views of the 4th ventricle and aqueduct were not completely satisfactory. There was very little air over the surface of the cerebral cortex.

Course. The child was discharged Nov. 26, 1954 with a diagnosis of hydrocephalus, most probably caused by failure of the subarachnoid channels to develop properly. The parents were instructed that pneumoencephalography was to be repeated in 1 month.

Readmission. The child was readmitted on Jan. 3, 1955 because of rapid enlargement of the head from 42 cm. to 47 cm. No other significant abnormality had been detected. Pneumoencephalography was repeated. This study revealed an obstruction in the 4th ventricle and poor filling of the lateral and 3rd ventricles. Air in the 4th ventricle seemed to outline a mass.

1st Operation. On Jan. 11, 1955 a suboccipital craniectomy was performed through a midline incision. Upon opening the dura mater, a smooth, white, glistening mass presented itself in the midline, approximately 3 cm. in diameter. A 25 gauge needle was inserted into the mass and arterial blood was obtained. Palpation of the mass then revealed an obvious pulsation and it was immediately apparent that a vascular malformation was present. Because of the limited exposure plus the fact that the origin of the vascular malformation was unknown, no definitive procedure was done and the operation was terminated.

Course. On Jan. 17, 1955 left vertebral angiography was performed with 35 per cent Diodrast. A large, midline aneurysmal sac was noted in the posterior fossa as had been seen at surgery (Fig. 1). It was thought that the feeding vessel was the left vertebral artery with drainage into the straight sinus.

2nd Operation. On Jan. 25, 1955 a suboccipital craniectomy was performed through a horseshoe-shaped incision. The aneurysmal mass was satisfactorily exposed. It was noted that the left vertebral artery divided before entering the large aneurysmal sac. Each division was clipped with an Olivecrona type of clip. No ill effects were noted and the aneurysm ceased to pulsate; however, this was only temporary and the aneurysm again started to pulsate in a few moments. The entire left cerebellar hemisphere was cystic and degenerative in nature, and the major portion of it was occupied by a huge angiomatous mass. The angiomatous mass together with the large aneurysm was dissected free and carefully divided from its vascular attachments. It was thus removed en bloc. After the removal, very little tissue remained of the left cerebellar hemisphere.

Course. The infant tolerated the procedure quite well. When discharged from the hospital his fontanelle had remained soft as well as the site of operation.
Pathologic Report. Gross. The fixed specimen is an irregularly shaped mass of tissue measuring approximately 4 cm. by 3.8 cm. by 3 cm. (Fig. 2). The main portion of this mass is made up of shaggy, dark-red tissue which has a soft, spongy consistency and is traversed by numerous small vascular spaces. Situated on one side of this mass of hemorrhagic tissue is a cystic structure which is round and measures 2.5 cm. in diameter. Several small vessels are seen entering this cyst, which has a smooth lining and a thin wall of rather firm, white tissue. It is filled with a clot of dark, red blood. There are five small openings into this cystic space, communicating with the hemorrhagic tissue outside. These openings are from 1 to 2 mm. in diameter.

Microscopic. Sections of the solid portion of the lesion reveal that it is composed predominantly of masses of small capillaries. Often these form ill-defined regions of small congested capillaries and are associated with some hemorrhage. Elsewhere they occur as rounded to ovoid, discrete masses containing similar capillaries. Most of the capillaries are quite well-formed and have small to slightly dilated lumens. They are lined by flat to slightly plump endothelium in which no mitotic figures were found after careful search. Only rarely small foci of endothelial cells with formed lumens are present. Portions of edematous cerebellar tissue are found frequently within the main mass of tumor. This cerebellar tissue is readily recognized, especially by portions of the molecular layer persisting. As a whole, there are relatively few macrophages present, but there is a moderate amount of hemorrhage.

Sections taken through the wall of the grossly described cyst reveal that it is made up of thickened muscular tissue which is somewhat edematous. It is lined by endothelium. The wall appears to be of uniform thickness and special stains fail to reveal any elastic tissue in it. These special stains confirm the presence of scattered, small, usually well-formed arteries, but these do not appear to be increased in number or abnormal in appearance. In a few places the capillary hemangiomatous elements are seen adjacent to the wall of the main cyst.

Because the main cystic mass lacks any elastic tissue in its wall, it is believed that this is derived from venous tissue. It is not clear just how this has been formed, but it is suspected that it is the result of the tumor creating an arteriovenous aneurysm, perhaps by destruction of the intervening tumor tissue.*

* Pathologic description by Dr. M. L. Trumbull, Chief, Department of Pathology, Baptist Memorial Hospital, Memphis, Tennessee.
Subsequent Course. The patient has been seen subsequently on a return visit to the clinic. The anterior fontanelle has remained soft and the head has not enlarged. The child has remained asymptomatic.

DISCUSSION

A careful search of the literature, although admittedly incomplete, has failed to reveal any case similar to this one in which an arteriovenous aneurysm of the posterior fossa has been successfully diagnosed and removed in an infant. The diagnosis in this case could have been made at the age of 5 weeks when the child apparently had his first spontaneous subarachnoid hemorrhage. This was evidenced by xanthochromia of the spinal fluid following his first seizure. Patterson in discussing the diagnosis of posterior fossa angiomas has emphasized that the symptomatology is almost identical to that produced by supratentorial angiomas. Cerebellar symptoms together with obstructive hydrocephalus suggest a tumor in the posterior fossa in most instances. The presence of subarachnoid hemorrhage suggests a vascular anomaly of the posterior fossa.

The pathologic report of the specimen describes the large aneurysmal sac, the wall of which is composed of muscular tissue. The fact that special stains failed to reveal any elastic tissue in the wall is significant and probably means that the sac is of venous origin. Surrounding the larger aneurysmal sac were masses of small capillaries, some of which were arterial. The stroma of the tumor mass was composed of edematous, cerebellar tissue. The microscopic picture of this specimen corresponds closely with that of a 16-year-old boy described by Olivecrona and Lysholm (Case 65). The tumor was exposed by a suboccipital approach. It appeared to be inoperable, the boy subsequently died from hemorrhage and at autopsy an angioma arteriale was proved to be present. As in our case, the vessels were separated by cerebellar tissue; however, the main vessel in their case was arterial as shown by the presence of elastic tissue in the wall with a well-formed elastica interna. The presence of cerebellar tissue between the vessels, as suggested by Cushing and Bailey, distinguishes the lesion as an arteriovenous aneurysm, as opposed to a true neoplasm, such as a hemangio-endothelioma of the cerebellum.

SUMMARY

A case report of a 2-month-old infant with an arteriovenous aneurysm of the posterior fossa has been described. The lesion was correctly diagnosed and treated surgically. No other case such as this has been noted in the literature.

REFERENCES