Super giant globoid intracranial aneurysm in an infant

Case report

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The incidence of intracranial aneurysms in infancy is less than 1%. Intracranial arterial aneurysms are very rare in patients under 1 year of age. This 6-month-old baby girl presented with progressive macrocephaly and anemia. Computerized tomography and cerebral angiography demonstrated a giant globoid aneurysm, 8 × 9 cm in size, arising from an anomalous posterior cerebral artery. The aneurysm was successfully clipped and excised in two stages. The child made an uneventful recovery. The congenital nature of the aneurysm is discussed, and the literature concerning intracranial aneurysms in patients under 1 year of age is reviewed.

KEY WORDS • giant aneurysm • infancy • congenital anomaly

Intracranial arterial aneurysms are very rare in childhood. In 1939, McDonald and Korb collected and reviewed 1125 cases of saccular intracranial aneurysms verified by operations or autopsy. Seventeen (1.5%) of these aneurysms occurred in children. Other published series from different neurosurgical centers reported a similar incidence of around 1%. Laitinen found a total of nine children among 688 patients with angiographically verified aneurysms, an incidence of 1.3%.

On the other hand, Housepian and Pool were unable to find any children among 113 cases of intracranial aneurysm collected over a 41-year period at the Columbia-Presbyterian Hospital in New York. In 1965, Matson published his series comprising 13 children with verified intracranial arterial aneurysms over a 12-year period, but his youngest patient was 16 months old. The youngest patient in McDonald and Korb's series was 18 months old. The youngest patient ever reported with a verified intracranial aneurysm was a boy who died at the age of 64 hours from the rupture of an aneurysm of the posterior communicating artery.

A review of the literature disclosed 15 cases in which the aneurysm was diagnosed during life in patients aged 1 year or younger. Of these patients, 11 were operated on successfully while four were considered inoperable, and three of those subsequently died (Table 1). Interestingly, the largest aneurysm in this group of patients measured 4 cm in diameter.

The youngest patient to be operated on was a 4-week-old girl reported by Jones and Shearburn in 1961, but the first successful operation after angiographic diagnosis was purportedly performed by Kimbell et al. in 1960, on a 16-month-old child.

We believe the intracranial aneurysm in the present case is the largest ever reported in an infant who underwent successful operation.

Case Report

This 6-month-old baby girl was admitted to the Children's Hospital of Eastern Ontario on June 20, 1979, after 4 weeks of progressive macrocephaly and 1 week of increasing irritability. One day prior to this admission, the patient started to vomit and refused to
feed. She had previously been in good health. Her perinatal history was unremarkable. Her developmental milestones were considered slightly delayed.

**Examination.** On physical examination, the infant was found very irritable and pale. Her occipitofrontal head circumference measured 47 cm, which exceeded the 98th percentile for her age. The anterior fontanel was large and bulging. Biparietal prominences were noted, with marked bulging on the left parietal region. Auscultation of the skull was negative for intracranial bruits, and cranial transillumination was negative. The hemoglobin on admission was 7.6 gm/dl, and the peripheral smear confirmed microcytic hypochromic anemia. In view of the history of macrocephaly and anemia, bilateral subdural taps were performed to rule out chronic subdural hematomas, and they were negative.

Shortly after admission, skull x-ray films demonstrated bulging and erosion of the left parietal bone, suggesting an expanding lesion in this area. A computerized tomography scan (Fig. 1) showed an enormous left hemispheric space-occupying lesion that was causing obstructive hydrocephalus. The lesion consisted of an area of increased density in the center, suggestive of clotted blood, surrounded by a well defined globular mass of contrast density, and between those two densities lay a narrow zone of radiolucency. This lesion was reported as a congenital neoplasm of uncertain origin; nevertheless, a lesion of vascular nature was suspected. Four-vessel selective cerebral angiography (Fig. 2) confirmed the presence of a very large arterial aneurysm, 8 X 9 cm in size, arising from an anomalous posterior cerebral artery. An intraluminal filling defect representing an intraluminal thrombus was also demonstrated.

**Operations.** On June 29, 1979, with the patient under general anesthesia, a Doppler probe was applied over the left parietal skull defect to continuously record the sounds from the intraluminal flow of the aneurysm during the procedure (Fig. 3). After this initial step and after the placement of an external ventricular drainage system, the aneurysm was ap-
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Fig. 3. Intraoperative Doppler recordings on the dome of the aneurysm. The arrow indicates the time of clipping. The fibrillation observed after clipping is artifact.

approached through a right subtemporal route, using magnification and microsurgical techniques. After retraction of the temporal lobe, the incisural region was reached. First the basilar artery was identified and then the anomalous right posterior cerebral artery was traced upward and forward toward its entrance into the aneurysmal sac. There was no clear aneurysm neck, and the parent vessel entered the sac directly. A Heifetz clip was placed across the parent vessel at approximately 1 cm from its entrance into the aneurysm, and the Doppler recording indicated the disappearance of the intracranial bruit (Fig. 3). A purse-string 4-0 silk suture was placed on the lateral wall of the aneurysm. The aneurysm sac was then punctured and aspirated, and at this stage bright red blood spurted out from the opening in the aneurysm, showing that the malformation was not completely excluded from the circulation. Controlled systemic hypotension of 40 torr was instituted for approximately 10 minutes. Further dissection of the parent vessel showed another feeding artery; this artery was clipped with a second Heifetz clip, and a third clip was placed at the junction of the parent vessel with the aneurysmal sac. Immediately after, the previously white sac became dark blue in color, indicating the total thrombosis of the aneurysm. A small opening of the wall was carried through the purse-string suture, and the presence of fresh intraluminal clot was confirmed. The purse-string suture was then closed. Total excision of the aneurysm was deemed impossible, and this first procedure was terminated. Postoperative angiography confirmed the successful clipping of the aneurysm (Fig. 4). Recovery from this procedure was uneventful, and postoperative CT

Fig. 4. Postoperative lateral and anteroposterior vertebral angiogram confirming the successful clipping of the aneurysm. The mass effect of the thrombosed aneurysm is still visible.
FIG. 5. Postoperative computerized tomography scan with contrast enhancement confirming the intraluminal thrombosis of the aneurysm after clipping. The preoperative obstructive hydrocephalus is no longer present after some shrinkage of the aneurysm; however, intracranial air, a right subdural effusion, and a subgaleal collection of blood are obvious. Note the thinning and erosion of the left parietal bone.

FIG. 6. Dramatic changes in the intracranial compartment are demonstrated by this computerized tomography scan with contrast enhancement, 6 months after resection of the aneurysm. Note the change in the skull shape as compared to Figs. 1 and 5.

demonstrated the total thrombosis of the aneurysm, with minimal shrinkage of its wall (Fig. 5). Because of the persistent mass effect of the thrombosed aneurysm, it was thought that the aneurysm should be excised. On July 17, 1979, the giant aneurysmal sac was approached through a left parietal craniotomy, and the dome of the aneurysm was exposed through a small parietal corticotomy. The dome of the aneurysm was opened, and 110 cc of organizing thrombus was withdrawn. The mouth of the feeding vessel was identified and cauterized with bipolar coagulation. Part of the aneurysmal sac was resected, and the rest of it underwent a natural plication by the expanding hemisphere.

Postoperative Course. The postoperative period was uneventful, and the child was discharged on August 2, 1979, neurologically intact.

She was readmitted on August 13, 1979, with a left subdural effusion under tension that required a left subdural peritoneal shunt. A follow-up CT demonstrated the disappearance of the aneurysm (Fig. 6). Since then she has progressed remarkably well and has caught up with her developmental milestones. She remains neurologically intact with a mild seizure disorder well controlled with anticonvulsants.

Discussion

Morley and Barr\textsuperscript{19} defined giant aneurysms as those larger than 2.5 cm in diameter. On the other hand, Sadik, \textit{et al.}\textsuperscript{20} stated that an aneurysm qualified for the denomination of "giant" if it was in the range of 7 × 5 × 5 cm. The aneurysm reported in this paper would fit this description of "giant." Obrador, \textit{et al.}\textsuperscript{21} reported the successful diagnosis and excision of a giant globoid aneurysm, 8 × 5 cm in size, arising from the posterior cerebral artery in a 20-year-old woman. Terao and Muraoka\textsuperscript{25} operated on a giant globoid aneurysm of the middle cerebral artery, 8 × 6 cm in size, in a 38-year-old patient; they postulated that such a giant aneurysm must be of a type different from the usual saccular aneurysm. Support for the congenital nature of such aneurysms was given in the presentation of a case from the records of the Massachusetts General Hospital,\textsuperscript{1} in which a globoid aneurysm of the right anterior cerebral artery, measuring 7 × 5 cm, was found in a 55-year-old man. It was believed that the globoid aneurysm represented a congenital anomaly and had less tendency to rupture than other types. There is little proof for this hypothesis, but the finding of such a giant globoid aneurysm in our infant patient provides definite evidence in support of this possibility. It would be hard to conceive how an ordinary saccular aneurysm could grow to this size as a consequence of repeated bleeding, organization, and accretion of clot over a period of years, or by simple dilatation of the aneurysmal wall. We must conclude, therefore, that the aneurysm was large from the beginning.

The ordinary saccular aneurysm owes its origin to a combination of developmental and degenerative changes involving the muscular coat and elastic layer of the vessel wall, as shown by Forbus,\textsuperscript{7} and corroborated by Richardson and Hyland\textsuperscript{22} and
Giant aneurysm in infancy

TABLE 1

Intracranial aneurysms diagnosed during life in early infancy

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Location of Aneurysm</th>
<th>Sex, Age</th>
<th>Clinical Presentation*</th>
<th>Surgical Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amacher &amp; Drake, 1975</td>
<td>middle cerebral artery</td>
<td>M, 12 mos</td>
<td>head injury</td>
<td>good</td>
</tr>
<tr>
<td>Bolander, et al., 1978</td>
<td>lt pericallosal artery</td>
<td>M, 6 mos</td>
<td>head injury, coma</td>
<td>fair</td>
</tr>
<tr>
<td>Ferry, et al., 1974</td>
<td>basilar artery</td>
<td>M, 3 mos</td>
<td>SAH, hydrocephalus, hemiparesis, ophthalmoplegia</td>
<td>no surgery; died at age 5 mos</td>
</tr>
<tr>
<td>Garcia-Chavez &amp; Moossy, 1965</td>
<td>lt middle cerebral artery</td>
<td>M, 5 mos</td>
<td>SAH</td>
<td>residual hemiparesis, seizures</td>
</tr>
<tr>
<td>Grode, et al., 1978</td>
<td>rt middle cerebral artery</td>
<td>F, 7 wks</td>
<td>SAH</td>
<td>excellent</td>
</tr>
<tr>
<td>Jones &amp; Shearburn, 1961</td>
<td>rt middle cerebral artery</td>
<td>F, 4 wks</td>
<td>convulsions, SAH</td>
<td>excellent</td>
</tr>
<tr>
<td>Lapras, et al., 1966</td>
<td>anterior choroidal artery</td>
<td>M, 11 mos</td>
<td>headache, vomiting</td>
<td>excellent</td>
</tr>
<tr>
<td>Lemmen &amp; Schneider, 1953</td>
<td>posterior cerebral artery</td>
<td>M, 8 mos</td>
<td>hydrocephalus</td>
<td>died</td>
</tr>
<tr>
<td>Morelli, et al., 1977</td>
<td>lt anterior cerebral artery</td>
<td>M, 4 mos</td>
<td>SAH</td>
<td>excellent</td>
</tr>
<tr>
<td>Shucart &amp; Wolpert, 1972</td>
<td>lt internal carotid artery</td>
<td>M, 6 mos</td>
<td>diabetes insipidus</td>
<td>excellent</td>
</tr>
<tr>
<td>Thompson, et al., 1973</td>
<td>lt posterior cerebral artery</td>
<td>F, 9 mos</td>
<td>cranial nerve, cerebellar signs</td>
<td>excellent</td>
</tr>
<tr>
<td>Thompson &amp; Pribram, 1969</td>
<td>rt internal carotid artery</td>
<td>F, 1 mo</td>
<td>ophthalmoplegia, hemiparesis</td>
<td>no surgery</td>
</tr>
<tr>
<td>Vapalahti, et al., 1969</td>
<td>rt middle cerebral artery</td>
<td>F, 3 mos</td>
<td>SAH, semicoma, hemiparesis</td>
<td>excellent</td>
</tr>
<tr>
<td>Ventureyra, et al., 1980</td>
<td>posterior cerebral artery</td>
<td>F, 6 mos</td>
<td>macrocephaly, anemia</td>
<td>excellent</td>
</tr>
</tbody>
</table>

*SAH = subarachnoid hemorrhage.
†The patient described by Lapras, et al. (1966) is reported by Laitinen.14

Carmichael. Bremer postulated that congenital aneurysms may arise in the following fashion. He suggested that, in the course of development of the arterial system of the brain, a plexus of small arteries from the base of the brain, almost all of which will atrophy and disappear, may sometimes and for unknown reasons remain, producing an aneurysmal sac that may undergo tremendous dilatation prenatally. It is conceivable that our patient may have developed her giant aneurysm in this manner.

Congenital giant globoid aneurysms in infancy, diagnosed during life, seem to be extremely rare. Review of the available literature (Table 1) indicates that our case may be unique regarding the patient's age, the size and location of the aneurysm, and the outcome after surgery. Jane reported a large aneurysm of the posterior inferior cerebellar artery, measuring 6 × 5 cm, that was found at autopsy in a 1-year-old child.

Giant globoid aneurysms of such dimensions as seen in our patient are apparently found more frequently in adults than in the pediatric age group, and this may indicate their silent evolution during the early years of life. The erosion of the skull in our patient strongly indicates that the aneurysm had been present for a considerable length of time, and it was asymptomatic until its bulk produced obstructive hydrocephalus. The sudden decompensation seems to be
related to partial intraluminal thrombosis, which also accounted for the patient’s anemia.

The recent technological advances in neurosurgery make the surgical treatment of such huge aneurysms in the pediatric age group feasible and safe.

References