Neonatal cavernous carotid artery aneurysm

Case report

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Neonatal intracranial aneurysms are rare. The authors report the case of a 4-week-old girl who presented with left-eye ptosis and proptosis. Computerized tomography scanning and magnetic resonance imaging demonstrated a mass involving the left cavernous sinus and middle cranial fossa. Cerebral angiography revealed a large complex left cavernous carotid artery (CA) aneurysm. The patient underwent endovascular treatment in which detachable coils and n-butyl cyanoacrylate glue were used to achieve complete obliteration of the aneurysm. To the authors' knowledge, this is the first reported neonatal intracranial aneurysm originating from the cavernous CA and treated endovascularly. The authors review the literature on neonatal intracranial aneurysms.

KEY WORDS • cavernous carotid artery • aneurysm • endovascular embolization • neonate • pediatric neurosurgery

Intracranial arterial aneurysms are uncommon in the pediatric population. In a cooperative study of intracranial aneurysms and SAH, only 41 (1.6%) of 2627 patients with aneurysmal SAH were younger than 19 years of age.16 In another large study involving 3000 ruptured intracranial aneurysms, only 58 lesions (1.9%) were present in patients younger than 19 years of age.21 In a different large series investigators reported on 50 patients (4.7%) younger than 20 years of age with ruptured saccular aneurysms of 1066 patients overall.24

Intracranial arterial aneurysms are rare in the neonatal period. Defining the neonatal period as the first 4 weeks of postnatal life, we identified only 17 cases in the literature. In this report, we describe a 4-week-old neonatal girl who presented with left-eye ptosis and proptosis and in whom a large cavernous CA aneurysm was diagnosed. We describe the clinical presentation, radiographic evaluation, endovascular treatment, and follow-up data obtained in this case and review the literature pertaining to this rare entity.

Abbreviations used in this paper: ACA = anterior cerebral artery; ACoA = anterior communicating artery; CA = carotid artery; CCA = common CA; CT = computerized tomography; ICA = internal CA; MCA = middle cerebral artery; MR = magnetic resonance; OphA = ophthalmic artery; PCoA = posterior communicating artery; PICA = posterior inferior cerebellar artery; SAH = subarachnoid hemorrhage; VA = vertebral artery.

Case Report

Presentation and History. This 4-week-old girl presented with a 1-week history of irritability and progressive left-eye ptosis and proptosis. Her birth had occurred after a full-term pregnancy in which she had been delivered via cesarean section performed secondary to fever and prolonged rupture of membranes in the mother. The Apgar scores were 9 and 9 at 1 and 5 minutes, respectively. Her perinatal history was remarkable for two failed left-ear otoacoustic hearing tests.

Examination. Physical examination revealed an awake, alert, and consolable neonate. Her occipitofrontal circumference was within the 90th percentile for her age. The anterior fontanel was soft and sunken. No cranial bruit was observed. The patient had left-eye proptosis and ptosis. Her pupils were equally round and reactive. Extraocular movements were normal on the right side; the patient had partial left-sided third and sixth cranial nerve palsies. Corneal reflexes were present bilaterally. On dilated funduscopic examination, the maculae were flat, and there were neither signs of venous congestion nor papilledema. The patient had a good cry, symmetrical Moro, and normal suck and grasp reflexes; muscle strength, tone, and bulk were normal.

Head CT scanning demonstrated a 1.8 × 1.6–cm hyperdense extraaxial mass in the left middle cranial fossa cen-
tered around the left cavernous sinus (Fig. 1). Erosive changes of the skull base were evident; there was enlargement of the foramen lacerum and spinosum, scalloping of the left side of the clivus, and erosion of the medial aspect of the petrous apex. There was no parenchymal or extraaxial hemorrhage. Brain MR imaging revealed an extraaxial mass in the left middle cranial fossa involving the left cavernous sinus (Fig. 2). The mass appeared heterogeneous on T1- and T2-weighted MR imaging sequences and heterogeneously enhanced following Gd administration. The mass encased the left ICA, and at the level of the cavernous sinus there was a 6-mm flow void contiguous with the ICA. There was mass effect both on the left temporal lobe and pituitary gland. The differential diagnosis included sarcoma, neuroblastoma, germ cell tumor, primitive neuroectodermal tumor, teratoma, histiocytosis, and aneurysm.

Angiography. Because of the uncertain origin of the mass, the patient underwent four-vessel cerebral angiography. Left CCA and ICA injections demonstrated delayed opacification of the left ICA, which ended at the skull base in a large complex aneurysmal lesion (Fig. 3). The aneurysm exhibited multiple compartments, and its largest pouch (measuring ~2 cm in diameter) was located lateral and superior to the cavernous sinus. There was opacification of the cavernous sinus and the superior ophthalmic veins bilaterally. The differential diagnosis included sarcoma, neuroblastoma, germ cell tumor, primitive neuroectodermal tumor, teratoma, histiocytosis, and aneurysm.

Endovascular Intervention. Because of the retrograde opacification of the aneurysmal lesion via the PCoA, left ICA occlusion alone was not believed to be effective in treating the lesion. Thus, transarterial detachable-coil embolization in combination with subsequent ICA occlusion was performed. A No. 5 French guiding catheter (MPC Envoy; Cordis Neurovascular, Miami, FL) was advanced into the left ICA. A No. 2.3 French microcatheter (Prowler Plus; Cordis Neurovascular) was then advanced over a No. 0.014 French guidewire (Transend 14; Boston Scientific Corp., Natick, MA) into the distal left ICA. Twenty-seven coils (J-coil, Detach 18; Cook, Inc., Bloomington, IN) were delivered under fluoroscopic control into the multiple aneurysmal compartments. Shorter coils were then used to occlude the distal left ICA (31 coils in all). Finally, 0.3 ml of NBCA glue (60% n-butyl cyanoacrylate, 40% ethiodol) (Trufill; Cordis Neurovascular) was injected through the coils in the ICA to ensure definitive long-term obliteration of the ICA.

Postprocedure control angiography of the left CCA revealed no flow in the distal left ICA or the aneurysmal lesion (Fig. 5). A right CCA angiogram demonstrated excellent collateral flow through the ACoA; a left VA injection revealed persistent flow within the OphA and no reflux into the aneurysmal lesion.

Postintervention Course. The patient experienced dramatic and rapid improvement in her proptosis. She did, however, exhibit a transient increase in cranial nerve palsies. Head CT scanning performed 2 days after embolization revealed no evidence of infarction. At the 5-month follow-up visit, the patient was developmentally normal with complete resolution of her left third nerve palsy and a resolving left sixth nerve palsy.

Discussion

Intracranial aneurysms are uncommon in the pediatric population. Although in some series focusing on pediatric aneurysms the authors have demonstrated some hetero-

**Fig. 1.** Left: Axial unenhanced CT scan demonstrating a hyperdense extraaxial mass in the left middle cranial fossa involving the left cavernous sinus. Right: Axial unenhanced bone-window CT scan demonstrating changes of the left skull base due to erosion.
geneity, several distinct characteristics have been elucidated. More specifically, compared with adult intracranial aneurysms, their pediatric counterparts are associated with a male predominance, predilection for the ICA bifurcation and the posterior circulation, and a higher incidence of giant aneurysms.6,20

Intracranial aneurysms rarely occur in the neonatal period. The first case was reported in 1949 by Newcomb and Munns,19 who described two neonates with intracranial aneurysms. Including that initial report, we have identified 18 cases, including our own, of neonatal intracranial aneurysms (Table 1).1,5,7,8,11–15,17,19,22,23,25,26,28
Several cases have been excluded in our analysis of neonatal intracranial aneurysms, including those reported in two embryos and in a stillborn fetus.\textsuperscript{2,30} Also excluded is a case reported by Thompson and Pribram\textsuperscript{27} of an infant girl in whom an ICA aneurysm had been diagnosed at 9 months of age and who experienced ophthalmoplegia and quadriplegia at 2 weeks of age. Her initial lumbar puncture at 1 month of age was negative. Grode, et al.,\textsuperscript{4} reported on a female newborn who suffered from lethargy, vomiting, and pallor on the 8th day of life and was treated for meningitis; an SAH was demonstrated at 5 weeks of life; and angiography revealed an MCA aneurysm at 7 weeks of life. This case was also excluded in the analysis of neonatal intracranial aneurysms.

Of the 18 reported cases, nine patients were female and nine were male. The most common presenting symptoms included irritability, vomiting, seizures, apnea, and cyanosis. Interestingly, our patient presented with proptosis and ptosis. In 17 of the 18 reports the aneurysm site was described. Eleven (65\%) of the 17 cases involved the anterior circulation). The most common site of origin was the MCA (seven cases [41\%]). Six (35\%) of the 17 cases occurred in the vertebrobasilar system. The second most common site of origin was the PICA (three cases [18\%]). In the present case, the cavernous CA was involved. To our knowledge, this is the first reported neonatal intracranial aneurysm arising from this intracranial vessel. The dimensions of the aneurysm sac were reported in only 16 cases:

![Preembolization angiograms](image1)

**FIG. 4.** Preembolization angiograms.  
A: Right CCA angiogram, anteroposterior view, demonstrating brisk collateral flow to the left ACA and MCA via the ACoA.  
B: Left VA angiogram, lateral view, demonstrating retrograde opacification of part of the aneurysmal lesion and opacification of the left OphA.

![Postembolization angiograms](image2)

**FIG. 5.** Postembolization angiograms.  
A: Left CCA angiogram, lateral view, demonstrating complete occlusion of the aneurysm after embolization and injection of glue.  
B: Right CCA angiogram, anteroposterior view, demonstrating collateral flow through the ACoA.  
C: Left VA angiogram, lateral view, demonstrating persistent flow within the OphA and no reflux into the aneurysmal lesion.
four (25%) were small (< 1 cm), 10 (62.5%) were large (1–2.5 cm), and two (12.5%) were giant (> 2.5 cm).

With only a handful of case reports of neonatal intracranial aneurysms, meaningful conclusions concerning the outcome of therapy are difficult to make. Nonetheless, several interesting observations can be made. In five cases the diagnosis was made after death.\textsuperscript{13,15,19,23} In the 13 patients in whom the diagnosis was established during life, one died without undergoing intervention,\textsuperscript{14} one was treated conservatively, 10 underwent surgery, and one (our case) underwent endovascular therapy. Interestingly, in the patient managed conservatively, the aneurysm was no longer detectable angiographically at 40 days of age.\textsuperscript{22} Of the 10 patients managed surgically, two died.\textsuperscript{17,26} In one case managed surgically, no further follow-up information is provided.\textsuperscript{1} In the remaining seven surgical cases in which follow-up data are provided, no neurological deficits were demonstrated.

In the present case, the patient was successfully treated with detachable coil placement. Although endovascular interventions have been used in the treatment of vein of Galen malformations,\textsuperscript{9,18} their application in the treatment of intracranial aneurysms in the very young has been reported in only a few cases.\textsuperscript{3,10,29} To our knowledge, the present case involves the youngest patient to have undergone coil embolization for an arterial intracranial aneurysm.

## References


## Table 1

Summary of cases of neonatal intracranial aneurysms reported in the literature\textsuperscript{*}

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age, Sex</th>
<th>Clinical Presentation</th>
<th>Site</th>
<th>Size</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Newcomb &amp; Munns, 1949</td>
<td>23 days, M</td>
<td>cyanosis &amp; tachypnea after mild head trauma cyanosis</td>
<td>circle of Willis</td>
<td>NR</td>
<td>no op</td>
<td>died</td>
</tr>
<tr>
<td></td>
<td>64 hrs, M</td>
<td>cyanosis</td>
<td>rt PCA–PCoA junction</td>
<td>2 mm</td>
<td>no op</td>
<td>died</td>
</tr>
<tr>
<td>Jones &amp; Shearburn, 1961</td>
<td>4 wks, F</td>
<td>crying, seizure</td>
<td>rt MCA</td>
<td>NR</td>
<td>clipped</td>
<td>no neurological abnormality died</td>
</tr>
<tr>
<td>Pickering, et al., 1970</td>
<td>1 mo, M</td>
<td>apnea, cyanosis, respiratory arrest, seizures hypotonia, esotropia, seizures, coma irritability, vomiting, apnea</td>
<td>rt PICA</td>
<td>2.5 × 1.2 × 2.5 cm</td>
<td>no op</td>
<td>died</td>
</tr>
<tr>
<td>Lee, et al., 1978</td>
<td>13 days, F</td>
<td>hypotonia, esotropia, seizures</td>
<td>BA</td>
<td>3 cm</td>
<td>no op</td>
<td>died</td>
</tr>
<tr>
<td>Lipper, et al., 1978</td>
<td>19 days, F</td>
<td>irritability, vomiting, apnea</td>
<td>lt ICA–PCoA junction</td>
<td>2.1 × 1.7 × 1.0 cm</td>
<td>ventric</td>
<td>died</td>
</tr>
<tr>
<td>Hungerford, et al., 1981</td>
<td>1 mo, F</td>
<td>crying, vomiting, pneumonia</td>
<td>rt MCA</td>
<td>1.5 × 1 cm</td>
<td>clipped</td>
<td>full recovery</td>
</tr>
<tr>
<td>Thrush &amp; Marano, 1988</td>
<td>1 mo, F</td>
<td>irritability, lethargy, agonal respirations</td>
<td>distal lt MCA</td>
<td>6 mm</td>
<td>clipped</td>
<td>no neurological deficit 20 days postop</td>
</tr>
<tr>
<td>Shimauchi, et al., 1989</td>
<td>19 days, F</td>
<td>vomiting, lethargy, seizure</td>
<td>branch of rt MCA</td>
<td>10 × 8 × 8 mm, fusiform</td>
<td>clipping of PA</td>
<td>no neurological deficits at 2.5 yrs</td>
</tr>
<tr>
<td>Piatt &amp; Clunie, 1992</td>
<td>18 hrs, M</td>
<td>seizures</td>
<td>distal lt SCA</td>
<td>1.5 cm</td>
<td>no op</td>
<td>well at 6 mos; normal angiogram at 40 days died</td>
</tr>
<tr>
<td>Kuchelmeister, et al., 1993</td>
<td>4 days, M</td>
<td>aspiration, apnea, seizures, hypotonia</td>
<td>ACoA</td>
<td>1 cm</td>
<td>no op</td>
<td></td>
</tr>
<tr>
<td>Hosotani, et al., 1995</td>
<td>24 days, M</td>
<td>hydrocephalus</td>
<td>distal lt PICA</td>
<td>15 mm</td>
<td>VP shunt, clipping of PA</td>
<td>normal neurological &amp; developmental findings at 7 mos alive/NR intraop cardiac arrest, died at 2wks</td>
</tr>
<tr>
<td>Allison, et al., 1998</td>
<td>1 mo, M</td>
<td>hydrocephalus</td>
<td>distal lt MCA</td>
<td>1 × 2 cm, fusiform</td>
<td>clipped</td>
<td>normal psychomotor development at 18 mos normal development at 1 yr</td>
</tr>
<tr>
<td>Jansen, et al., 2000</td>
<td>11 days, M</td>
<td>hydrocephalus, vomiting, reduced UO, respiratory failure</td>
<td>distal lt MCA</td>
<td>10 mm, fusiform</td>
<td>clipping</td>
<td>normal psychomotor development at 18 mos normal development at 1 yr</td>
</tr>
<tr>
<td>Kourtopoulos, et al., 2000</td>
<td>13 days, F</td>
<td>head &amp; eye deviation headache, decreased LOC, FITT</td>
<td>distal lt MCA</td>
<td>8 × 6 mm</td>
<td>clipping, VP shunt clipped (both)</td>
<td>no neurological deficit at 2 mos</td>
</tr>
<tr>
<td>Maroun, et al., 2003</td>
<td>3 days, M</td>
<td>irritability, cyanosis, seizures</td>
<td>distal lt MCA</td>
<td>2–3 mm</td>
<td>NR</td>
<td></td>
</tr>
<tr>
<td>present case</td>
<td>1 mo, F</td>
<td>irritability, cyanosis, seizures</td>
<td>cavernous CA</td>
<td>2 cm</td>
<td>coil embolization</td>
<td>intraop cardiac arrest, PVS, died at 4 yrs resolving 6th nerve palsy at 5 mos</td>
</tr>
</tbody>
</table>

\* BA = basilar artery; FTT = failure to thrive; LOC = level of consciousness; NR = not reported; PA = parent artery; PCA = posterior cerebral artery; PVS = persistent vegetative state; SCA = superior cerebellar artery; UO = urine output; ventric = ventriculostomy; VP = ventriculoperitoneal.
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