Idiopathic distal lenticulostriate artery aneurysm in a child

Case report and review of the literature

HAMILTON MATUSHITA, M.D., PH.D., ROBISON LUIS OLIVEIRA AMORIM, M.D.,
WELLINGSON SILVA PAIVA, M.D., DANIEL DANTE CARDEAL, M.D.,
AND FERNANDO CAMPOS GOMES PINTO, M.D.

Pediatric Neurosurgery, Department of Neurosurgery, University of São Paulo Medical School, São Paulo, Brazil

✓ The authors describe a rare case of idiopathic distal lenticulostriate artery (LSA) aneurysm in a 5-year-old boy who presented in the emergency department with a sudden onset of headache. Admission computed tomography scans revealed an intracerebral hemorrhage in the left caudate nucleus with intraventricular extension. Angiographic studies demonstrated a left medial LSA aneurysm. The patient underwent a left parasagittal frontal craniotomy, the lateral ventricle was accessed via the anterior transcallosal approach, and the aneurysm was removed after sectioning of the parent vessel. The child left the hospital after 5 days; at that time he was asymptomatic and without motor impairment. The optimum treatment of aneurysms involving small perforating arteries is controversial and depends mainly on the causative factors. The pathogenesis and treatment of these unusual aneurysms are discussed.

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KEY WORDS • cerebral aneurysm • children • distal aneurysm • lenticulostriate artery • pediatric neurosurgery

A NEURYSMS occurring on small, distal, and straight segments of cerebral arteries are extremely uncommon. In a survey of the literature we found only 22 cases of aneurysms of the distal segment of the LSA, three of them in children. We describe a distal medial LSA aneurysm in a 5-year-old child with ganglionic hemorrhage not related to other diseases. These aneurysms are embedded in the deep basal brain parenchyma, not in the subarachnoid or cisternal spaces, which makes them surgically inaccessible (our case was a very unusual one, in which the hematoma cavity provided access). The reported case was approached via the lateral ventricle, through the hematoma cavity, and the aneurysm was removed after sectioning of the parent vessel without causing neurological deficits.

Case Report

History and Examination. This 5-year-old boy presented in our emergency department with a history of sudden onset of headache during sleep, causing him to wake up, followed by vomiting and sleepiness, without loss of consciousness. He had been healthy with no constitutional or connective tissue disorders and no infectious or neurological symptoms. On admission he was alert and interacting but irritated, and no signs of motor deficits were identified. Admission CT scans revealed an intracerebral hemorrhage in the left caudate nucleus, predominantly involving the head and body of the nucleus, with rupture into the anterior horn of the lateral ventricle and moderate hydrocephalus (Fig. 1).

Initial Treatment. The patient was treated conservatively in an intensive care unit and exhibited mild headache, confusion, and somnolence for the first 3 days. A digital subtraction angiography study of the brain was performed and demonstrated an aneurysm 4 mm in diameter arising from the distal portion of the left medial LSA (Fig. 2). Vasospasm was not observed, and the left carotid and vertebral arteries demonstrated no abnormalities. These radiological findings indicated that the hemorrhage was due to aneurysm rupture. Conservative therapy was chosen, with close observation of the course of the hematoma and of the neurological status of the child. Serial CT scans were obtained to observe the evolution of the ventricular dilation, but no ventricular drainage was necessary. Two weeks later the angiography study was repeated, and there were no significant differences compared with the earlier one. Attempts to catheterize the LSA failed due to the acute angle formed by the origin of the vessel and the anterior cerebral artery. A transthoracic Doppler echocardiography study was negative for endocarditis. No other sources of infection, such as menin-
gitis, sinusitis, or skull osteomyelitis were detected. Laboratory examination and blood cultures failed to identify a pathogen and no blood dyscrasias were demonstrated.

Operation. After 15 days the patient underwent a left frontal parasagittal craniotomy. The lesion was approached via the interhemispheric anterior transcallosal route. There was a hematoma in the lateral ventricular cavity, and a rupture of the ependymal lining at the lateral and inferior wall of the lateral ventricle, just at the head of the caudate nucleus. The intraparenchymal part of the hematoma was removed through this opening. The aneurysm was clearly found to be involved in a blood clot at the wall of the posterior part of the hematoma cavity. The aneurysm was partly thrombosed and had to be removed after sectioning the medial LSA. The child recovered very quickly in the operating room with no motor deficit, and was sent to the intensive care unit. Postoperatively, there was no worsening of the patient’s neurological status.

Postoperative Course. A postoperative CT scan demonstrated complete removal of the cerebral hematoma (Fig. 3). The child had an uneventful postoperative recovery and was discharged from the hospital after 5 days, with no neurological deficit. Microscopic examination demonstrated a 4-mm-diameter aneurysm that was partially occluded by a firm dark clot, with a very thin arterial wall and no inflammatory reaction.

Discussion

Diagnostic Criteria

Intracranial aneurysms are rarely found in childhood and adolescence, although they constitute the most frequent cause of intracranial bleeding at this age. A total of 706 cases of pediatric intracranial aneurysm have been described in the literature since 1939. Aneurysms in infancy and childhood correspond to between 1 and 3% of all intracranial aneurysms. One of the differences that has been reported between adult and childhood aneurysms is their location. A high incidence of pediatric aneurysms at the internal carotid artery bifurcation and in the posterior circulation has been reported by many authors. Aneurysms in infancy and childhood correspond to between 1 and 3% of all intracranial aneurysms.

For the distal portion of the LSA to be the site of an aneurysm is very unusual. A review of the literature shows 19 reported cases in adults and only three in children (Tables 1 and 2). All of the pediatric cases, including our patient, presented with basal ganglia and intraventricular hemorrhage. The majority of LSA aneurysms have been considered to be idiopathic. Causative factors related to LSA aneurysms as identified from the literature are as follows: moyamoya disease, rheumatic diseases, arteriovenous malformation, and intraventricular tumor.

Aneurysm formations in moyamoya disease are more frequent in the collateral vessels, either in the perforating arteries or in the posterior circulation. Approximately 40% of the associated aneurysms are located at the peripheral arteries, mainly at the distal portion of the perforating and choroidal arteries. The mechanism of formation of the peripheral aneurysms may be related to hemodynamic stress on the moyamoya vessels, which are dilated to compensate for the occlusive change in the main trunk of the anterior circulation of the circle of Willis. Kodama and Suzuki reported three cases of moyamoya disease associated with peripheral aneurysms that disappeared on the follow-up angiogram, suggesting that aneurysms associated with moyamoya disease may be pseudoaneurysms resulting from...
previous ruptures of these small arteries. It is controversial whether the deep hemorrhages in moyamoya disease are secondary to unrecognized microaneurysms or to the rupture of dilated fragile moyamoya vessels. The same pathogenetic mechanism of hemodynamic stress due to increased demand for blood supply to the tumor was found in the case of an LSA aneurysm associated with intraventricular neurocytoma.

Vasculitis associated with distal cerebral aneurysms is rarely reported in the literature, although its role in the origin of cerebral aneurysms has not been clarified. Sakaki et al. reported on a 29-year-old woman with systemic lupus erythematosus who had a ruptured cerebral aneurysm at the trifurcation of the MCA. Asaoka et al. described the case of a 54-year-old woman with aortitis who had unruptured cerebral aneurysms in the left internal carotid and anterior communicating arteries. Nakasu et al. reported on a 47-year-old man with Behçet disease who presented with a subarachnoid hemorrhage due to rupture of a distal MCA aneurysm. Heran et al. described the case of a 43-year-old woman with isolated primary angiitis of the central nervous system in whom multiple aneurysms of the LSA were found.

It is particularly important to rule out infectious disease as the cause of small and peripheral cerebral artery aneurysms. The distal MCA is the most common site of origin of septic cerebral aneurysms, and bleeding into the brain parenchyma is typically common due to the location of these lesions distally on vessels deep within the cerebral sulci or perforating arteries. Infectious intracranial aneurysms comprise 2 to 5% of all intracranial aneurysms and children are more commonly affected, particularly children with congenital heart disease, artificial heart valves, rheumatic heart disease, or acquired bacterial endocarditis. No case of infectious aneurysms affecting the LSA has been reported (Tables 1 and 2).

### Treatment Options

Surgical management is controversial in distal cerebral aneurysms associated with moyamoya disease and in infectious cerebral aneurysms. If the lesion is associated with moyamoya disease, Kuroda et al. recommend direct bypass surgery because it can reduce the hemodynamic stress through the collateral vessels; the blood flow through the external carotid artery system increases after such an operation. These authors pointed out that rebleeding occurs in approximately 20% of cases and that preventive surgery should be performed to deal with such a high incidence of rebleeding. Kawaguchi et al. reported that peripheral artery aneurysms, which are related to moyamoya disease, may rupture more than twice, although the lesions may disappear spontaneously between 4 and 60 months (mean 7.1 months) after the initial presentation. Direct surgery of the aneurysm and cerebral revascularization performed at the same time are proposed by Hamada et al. for patients with repeated hemorrhages. Direct surgery may carry a risk of additional neurological deficits because peripheral aneurysms usually are located in the eloquent areas or periventricular deep brain tissue. Others propose conservative treatment and repeated angiography, because many aneurysms involving small arteries tend to disappear spontaneously.

The optimum treatment for infectious intracranial aneurysms is surrounded by many controversies, and there are multiple treatment modalities available currently; these consist of medical, surgical, and endovascular therapies. Treatment with antibiotics and serial angiographic studies is generally advised for proximal arterial trunks, unruptured aneurysms, multiple lesions, or aneurysms involving arteries in eloquent areas of the brain. The mechanism proposed for the observed spontaneous thrombosis of infectious aneurysms implicates the inflammatory process aiding the thrombosis and repair. Surgery should be reserved for

### TABLE 1

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age, Sex</th>
<th>CT Findings</th>
<th>Origin</th>
<th>Treatment</th>
<th>Pathology</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Endo et al., 1996</td>
<td>12 yrs, F</td>
<td>ICH + IVH</td>
<td>idiopathic</td>
<td>proximal clipping</td>
<td>not performed</td>
<td>good</td>
</tr>
<tr>
<td>Kaptain et al., 2001</td>
<td>2 mos, M</td>
<td>ICH + IVH</td>
<td>idiopathic</td>
<td>extirpation</td>
<td>true aneurysm</td>
<td>good</td>
</tr>
<tr>
<td>Lehmann et al., 2003</td>
<td>2 yrs, F</td>
<td>IVH</td>
<td>idiopathic</td>
<td>extirpation</td>
<td>true aneurysm</td>
<td>NR</td>
</tr>
<tr>
<td>present study</td>
<td>5 yrs, M</td>
<td>ICH + IVH</td>
<td>idiopathic</td>
<td>extirpation</td>
<td>true aneurysm</td>
<td>good</td>
</tr>
</tbody>
</table>

* ICH = intracerebral hemorrhage; IVH = intraventricular hemorrhage; NR = not reported.
patients in whom the aneurysm is seen to enlarge on follow-up angiography, or if the lesion does not decrease after the patient receives the antibiotic course. Bohmfalk et al. observed that the mortality rate in the nonsurgical group was considerable, with death occurring in 53%. Frazee et al. reported a high mortality rate for patients with nonsurgically treated aneurysms and advocated excision. Ojemann recommended direct surgery for accessible aneurysms on the distal branch of the MCA associated with intracranial hemorrhage. The surgeon should be prepared to use different operative strategies in dealing with various situations that can arise in patients with infectious intracranial aneurysms.

No causative factor was associated with our case, and therefore it was considered to be an idiopathic case of distal cerebral artery aneurysm. In this situation, the natural history of the aneurysm is unclear and treatment should be individualized. There is no doubt that whenever an acutely life-threatening intracerebral hematoma occurs, it should be removed and the aneurysm should be treated at the same time.

Direct surgical treatment of aneurysms affecting small perforating vessels is not recommended because this type of lesion can be a pseudoaneurysm, and a second angiographic study may reveal occlusion of the aneurysm. In three reported cases the aneurysm disappeared without surgical treatment. Although the risk of rebleeding was unknown, and the natural course of untreated intracranial aneurysms is toward progressive weakening of the aneurysm wall, we opted for surgical treatment, approaching through the cavity of the hematoma. Because this aneurysm was located in the brain tissue around the lateral and inferior portion of the lateral ventricle, the hematoma provided an opportunity to reach the lesion without opening the deep brain tissue. Schürmann et al. described a similar case in which the aneurysm appeared on the surface of the lateral ventricle, and which they approached through a cortical incision.

We chose a transcallosal approach because of the small size of the lateral ventricles. This route provided excellent visualization of relevant anatomical landmarks within the lateral ventricle and the site of ependymal lining rupture by the basal ganglia hematoma. Without the hematoma as a guide, it would be difficult to localize the aneurysm in the basal ganglia; therefore, it is highly advisable to perform the surgery within 2 weeks of the hemorrhage. The main justification for the delay of 15 days before surgical treatment was performed in our case was the fact that we initially accepted the possibility of spontaneous obliteration of this small, distal, deep aneurysm. Another point of concern is related to preservation of the parent artery. Proximal ligation of the LSA through the sylvian fissure and basal cistern might be more hazardous due to the difficulty in identifying the precise parent artery, and proximal occlusion of the artery could result in larger areas of ischemia than distal ligation. Peripheral aneurysms are usually fusiform, without a neck, with irregular and eccentric profiles that suggest pseudoaneurysms, or as in the case of infectious intracranial

### TABLE 2

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs.), Sex</th>
<th>CT Findings</th>
<th>Origin</th>
<th>Treatment</th>
<th>Pathology</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Schürmann et al., 1968</td>
<td>23, F</td>
<td>ICH + IVH</td>
<td>idiopathic</td>
<td>extirpation</td>
<td>true aneurysm</td>
<td>good; no motor deficit</td>
</tr>
<tr>
<td>Ohta et al., 1980</td>
<td>26, M</td>
<td>ICH + IVH</td>
<td>AVM</td>
<td>extirpation</td>
<td>true aneurysm</td>
<td>vegetative</td>
</tr>
<tr>
<td>Okuma et al., 1980</td>
<td>29, F</td>
<td>not performed</td>
<td>moyamoya</td>
<td>spontaneous regression</td>
<td>not performed</td>
<td>good</td>
</tr>
<tr>
<td>Murakami et al., 1984</td>
<td>33, M</td>
<td>ICH + IVH</td>
<td>moyamoya</td>
<td>none</td>
<td>true aneurysm</td>
<td>died of re-bleeding</td>
</tr>
<tr>
<td>Kidoguchi et al., 1987</td>
<td>24, F</td>
<td>SAH + ICH</td>
<td>lupus erythematosis</td>
<td>extirpation &amp; conservative</td>
<td>true aneurysm</td>
<td>not performed</td>
</tr>
<tr>
<td>Grabel et al., 1989</td>
<td>60, M</td>
<td>ICH</td>
<td>moyamoya</td>
<td>drainage of hematoma</td>
<td>true aneurysm</td>
<td>died</td>
</tr>
<tr>
<td>Gupta et al., 1989</td>
<td>36, F</td>
<td>ICH</td>
<td>idiopathic</td>
<td>conservative</td>
<td>not performed</td>
<td>good</td>
</tr>
<tr>
<td>Oka et al., 1991</td>
<td>44, F</td>
<td>ICH</td>
<td>1 hypertension, 1 moyamoya</td>
<td>conservative</td>
<td>not performed</td>
<td>good</td>
</tr>
<tr>
<td>Petrela et al., 1992</td>
<td>32, M</td>
<td>ICH</td>
<td>idiopathic</td>
<td>extirpation</td>
<td>true aneurysm</td>
<td>motor deficit</td>
</tr>
<tr>
<td>Kuroda et al., 2001</td>
<td>27, M</td>
<td>ICH</td>
<td>moyamoya</td>
<td>bypass surgery</td>
<td>not performed</td>
<td>good</td>
</tr>
<tr>
<td>Larrazabal et al., 2001</td>
<td>50, F</td>
<td>IVH (3rd ventricle)</td>
<td>moyamoya</td>
<td>bypass surgery</td>
<td>not performed</td>
<td>good</td>
</tr>
<tr>
<td>Vates et al., 2001</td>
<td>57, F</td>
<td>ICH + IVH</td>
<td>MCA occlusion</td>
<td>endovascular (NBCA)</td>
<td>not performed</td>
<td>good</td>
</tr>
<tr>
<td>Heran et al., 2003</td>
<td>43, F</td>
<td>ICH</td>
<td>arteritis</td>
<td>immunosuppressive therapy</td>
<td>not performed</td>
<td>good</td>
</tr>
<tr>
<td>Lehmann et al., 2003</td>
<td>26, M, 26, M, 26, F</td>
<td>IVH</td>
<td>2 idiopathic, 1 moyamoya</td>
<td>conservative</td>
<td>not performed</td>
<td>2 good, 1 satisfactory</td>
</tr>
<tr>
<td>Narayan et al., 2004</td>
<td>69, F</td>
<td>ICH + IVH</td>
<td>idiopathic</td>
<td>arterial ligation (cisternal approach)</td>
<td>not performed</td>
<td>good</td>
</tr>
<tr>
<td>Sakai et al., 2005</td>
<td>61, M</td>
<td>ICH</td>
<td>moyamoya</td>
<td>neck clipping of aneurysm through hematoma cavity</td>
<td>not performed</td>
<td>good</td>
</tr>
</tbody>
</table>

* AVM = arteriovenous malformation; NBCA = N-butyl cyanoacrylate; SAH = subarachnoid hemorrhage.
aneurysms, appear with destruction of the artery wall, with its weakness and friability. Treatment of these aneurysms conventionally with clip occlusion and preservation of the parent vessels is the exception. A dissecting type of aneurysm was a possibility in our case, due to its irregular profile, diffuse neck, and location in a straight portion of the artery. A defect of unknown origin in the internal elastic lamina may result in dissection of the vessel layers, aneurysmal dilation of the artery, and accumulation of blood within its layers, with stenosis of its lumen.

Endovascular management with coil placement in such a friable and thin-walled aneurysm is potentially hazardous. Endovascular treatment of peripheral aneurysms is likely to involve parent vessel sacrifice, so this therapeutic modality should be indicated in noneloquent areas of the brain. In addition, insertion of the guidewire into such a thin artery is likely to be difficult. Early recognition and treatment of this source of basal ganglia bleeding avoids a major neurological deficit or death caused by rebleeding of the aneurysm. As shown in Tables 1 and 2, the majority of patients with LSA aneurysms may have a good prognosis, and this is unrelated to either the origin of the lesion or the treatment modality.

Conclusions

This unusual case, and the review of the literature, demonstrate the rarity of the LSA as a site for an aneurysm and that the pathophysiological basis for development of aneurysms affecting small and distal arteries must be different from that of the usual saccular lesions arising at a branching site on a major proximal artery. As our review of the literature indicated, many cases involving aneurysms of LSAs have an associated pathological entity. In the cases classified as idiopathic, including ours, the findings lead to the conjecture that a defect of the arterial wall results in dissection of the adventitia and formation of an aneurysm. Review of the literature indicated a varied modality of treatment. For idiopathic cases, it seems that surgical treatment of the aneurysm is the option of choice, although the risk of rebleeding is unknown and some patients treated conservatively achieved good results.

References


38. Petrela M, Xhurmari A, Azdurian E, Vreto G: [Aneurysm of the terminal part of the lenticulostriate artery.] Neurochirurgie 38:50–52, 1992 (Fr)