Intracranial giant aneurysms in children and adolescents misdiagnosed as intracranial tumors before operation: 2 cases report

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Abstract: The incidence of giant intracranial aneurysms in the pediatric population is rare, they are prone to contain thrombi and look like tumors in CT or MRI scans. Our report presents two fully thrombosed giant, middle cerebral artery aneurysms mimicking intracranial tumors in a 16 month old pediatric patient and a 17 year old girl before operation. Surgical clipping and vascular anastomosis were performed to these two patients and they were recovered with no neurological deficits in six month’s follow-up. Our cases show that giant thrombosed aneurysms can be potentially misdiagnosed as neoplasms in children, great care must be exercised when managing such cases, and surgical treatment of giant pediatric aneurysms can get excellent outcomes.

Keywords: Giant aneurysms, children, germ cell tumor, glioma, aneurysm occlusion

Introduction

Intracranial aneurysms (IA) in children are rare, occupying 0.17% to 4.6% of the total cases of this disease in the general population [1-5]. Pediatric intracranial aneurysms represent a pathophysiological entity different from their adult counterparts, which are not simply “standard” saccular aneurysms and usually have a higher incidence of giant or complex aneurysms with a huge intracranial space occupying effect. Thus they may be misdiagnosed as tumors before operation. In addition, they require more challenging treatment strategies like intracranial bypass procedures or therapeutic parent artery occlusion. Here we report two cases of pediatric intracranial giant aneurysms that were misdiagnosed as tumors before surgery to reference for counterparts.

Case report

Case 1

Due to a sudden right upper limb twitch for a few seconds, a 16 month old patient underwent an emergency CT scan in a local hospital.
treatment. After opening the dura, we saw the local frontal and temporal lobe brain tissue was soft and yellow, and contained multiple brown cysts. After releasing the sac, there was a 5 cm × 4 cm mass in the sac cavity with a clear boundary. The cyst was complete and the quality was hard. After exposing the lateral fissure we found the upper branch of the middle cerebral artery directly transferred into the aneurysm sac, and the distal end of the artery was directly transferred into the superior sagittal sinus to form an arteriovenous fistula. We explored around the aneurysm wall and found no vascular supply to the surrounding brain tissue from the aneurysm. We then clipped the proximal end of the artery to cut off the blood supply to the aneurysm to reduce tension, and then we cut off the distal end of the aneurysm artery and separated it to complete resection of the aneurysm (Figure 2).

Six months later, the follow-up showed that the patient recovered well with no nervous system defects. Right upper limb muscle activity was
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Case 1

A 5 year old female had a history of headaches and vomiting for one week. The patient received a CT and DSA review and no abnormal characteristics were found. Three years later, the patient received a CT and DSA review and no abnormal characteristics were found.

Case 2

An 18 year old female suffered a blunt headache for one week. The local hospital diagnosed a right temporal lobe tumor using MRI. For further treatment, the patient was transferred to our hospital. After admission the patient presented no positive signs except for headache. We gave her an MRI scan again and a T1 image showed a flake low signal mass with hyperintensity occupying the posterior part of the right temporal lobe. A T2 image showed a mixed flake low signal and high signal. Brain tissue edema was obvious and the right ventricle restored to normal. Three years later, the patient received a CT and DSA review and no abnormal characteristics were found.

Figure 3. A, B. A T1 image showed a flake low signal mass with hyperintensity in the posterior part of the occupying in the right temporal lobe. A T2 image showed a mixed flake low signal and high signal. C, D. An enhanced scan showed the front of a low signal mass in T1.

Figure 4. A, B. The aneurysm was separated with two temporary blocking clips clipping both ends of it. The aneurysm was completely resected and the proximal and distal ends of the artery were freed for end-end anastomosis. C. The size of the aneurysm was 4.5 cm × 3 cm. D. A CTA review showed middle cerebral artery patency. The arrow indicates the site of anastomosis. E, F. The MRI review after 3 months showed no abnormalities.
was severely compressed. An enhanced T1 scan showed the front of a low signal mass and we misdiagnosed it as a glioma with tumor apoplexy (Figure 3).

The patient then received routine examinations and had no surgical contraindications. We chose a right temporal flap craniotomy approach to dissect the tumor. After incising the dura mater we saw part of the brain tissue of the temporal lobe was yellow. About 5 mm below the yellow portion of the cortex we found a pulsatile mass with smooth and clear borders. It was a giant aneurysm (Figure 4). The size of the mass was 4.5 cm × 3 cm × 2 cm and the surrounding brain tissue contained a hemosiderin deposition. After the aneurysm was carefully separated, the proximal end of the aneurysm was found to be connected to the M2 segment of the middle cerebral artery and the distal end to the temporal occipital artery. Both ends of the aneurysm were clipped with two temporary blocking clips. Then the aneurysm was completely resected and the proximal

Table 1. Summary of the literature between 2000 and 2017 reporting the giant cerebral aneurysms in a pediatric population

<table>
<thead>
<tr>
<th>Year</th>
<th>Authors</th>
<th>Giant aneurysm Patients</th>
<th>Male/Female</th>
<th>Treatment</th>
<th>Outcome (GOS)</th>
</tr>
</thead>
<tbody>
<tr>
<td>2000</td>
<td>Dorfler et al [23]</td>
<td>1</td>
<td>0/1</td>
<td>Coiling</td>
<td>4 (visual loss)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1 GOS 3</td>
</tr>
<tr>
<td>2001</td>
<td>Desai et al [24]</td>
<td>1</td>
<td>0/1</td>
<td>Conservative</td>
<td>1</td>
</tr>
<tr>
<td>2001</td>
<td>Otsuka et al [25]</td>
<td>1</td>
<td>1/0</td>
<td>Craniotomy</td>
<td>3</td>
</tr>
<tr>
<td>2002</td>
<td>Zhang et al [15]</td>
<td>2</td>
<td>1/1</td>
<td>Craniotomy</td>
<td>1 GOS 5</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1 GOS 4</td>
</tr>
<tr>
<td>2002</td>
<td>Krapf et al [26]</td>
<td>1</td>
<td>0/1</td>
<td>Conservative</td>
<td>5</td>
</tr>
<tr>
<td>2003</td>
<td>Massimi et al [27]</td>
<td>1</td>
<td>0/1</td>
<td>Coiling</td>
<td>1</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>4 Conservative 1</td>
<td>1 GOS 4</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>1 GOS 1</td>
</tr>
<tr>
<td>2005</td>
<td>Krishna et al [28]</td>
<td>1</td>
<td>1/0</td>
<td>Conservative</td>
<td>1</td>
</tr>
<tr>
<td>2006</td>
<td>Jurkiewicz et a [29]</td>
<td>1</td>
<td>1/0</td>
<td>Craniotomy</td>
<td>4</td>
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<tr>
<td>2007</td>
<td>Khan et al [30]</td>
<td>1</td>
<td>1/0</td>
<td>Craniotomy</td>
<td>5</td>
</tr>
<tr>
<td>2008</td>
<td>Kasiwal et al [31]</td>
<td>1</td>
<td>1/0</td>
<td>Conservative</td>
<td>5</td>
</tr>
<tr>
<td>2008</td>
<td>Malhotra et al [32]</td>
<td>1</td>
<td>0/1</td>
<td>Conservative</td>
<td>1</td>
</tr>
<tr>
<td>2009</td>
<td>Goedee et al [33]</td>
<td>1</td>
<td>0/1</td>
<td>Craniotomy</td>
<td>5</td>
</tr>
<tr>
<td>2009</td>
<td>Kantarci et al [34]</td>
<td>1</td>
<td>0/1</td>
<td>Conservative</td>
<td>1</td>
</tr>
<tr>
<td>2010</td>
<td>Wilsms et al [35]</td>
<td>1</td>
<td>0/1</td>
<td>Coiling</td>
<td>3</td>
</tr>
<tr>
<td>2010</td>
<td>Rehman [36]</td>
<td>1</td>
<td>1/0</td>
<td>Craniotomy</td>
<td>5</td>
</tr>
<tr>
<td>2010</td>
<td>Zomorodi et al [37]</td>
<td>1</td>
<td>1/0</td>
<td>Craniotomy</td>
<td>5</td>
</tr>
<tr>
<td>2012</td>
<td>Lan et al [38]</td>
<td>1</td>
<td>1/0</td>
<td>Craniotomy</td>
<td>4</td>
</tr>
<tr>
<td>2012</td>
<td>Al Youbi et al [39]</td>
<td>1</td>
<td>0/1</td>
<td>Craniotomy</td>
<td>5</td>
</tr>
<tr>
<td>2013</td>
<td>Mrak G et al [22]</td>
<td>1</td>
<td>0/1</td>
<td>Craniotomy</td>
<td>5</td>
</tr>
<tr>
<td>2014</td>
<td>Goncalves et al [40]</td>
<td>1</td>
<td>1/0</td>
<td>Conservative</td>
<td>4</td>
</tr>
<tr>
<td>2015</td>
<td>Fathi NQ et al [20]</td>
<td>1</td>
<td>1/0</td>
<td>Craniotomy</td>
<td>5</td>
</tr>
<tr>
<td>2016</td>
<td>Demartini et al [41]</td>
<td>1</td>
<td>1/0</td>
<td>Coiling</td>
<td>5</td>
</tr>
<tr>
<td>2016</td>
<td>De Aguial et al [42]</td>
<td>1</td>
<td>1/0</td>
<td>Conservative</td>
<td>4</td>
</tr>
<tr>
<td>Total</td>
<td></td>
<td>33</td>
<td>17/16</td>
<td>Conservative 10</td>
<td>16 GOS 5</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Craniotomy 17</td>
<td>7 GOS 4</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Coiling 6</td>
<td>3 GOS 3</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>6 GOS 1</td>
</tr>
</tbody>
</table>
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and distal ends of the artery were freed for an end-end anastomosis.

A computed tomography angiography (CTA) review showed middle cerebral artery patency. Three months later, the patient was followed up with. No nervous system defects were detected, muscle strength was normal, and the MRI review showed no abnormalities.

Discussion

Intracranial aneurysms in children (<18 years old) represent between 0.17 and 4.6% of the total number of the disease in the general population [1-5]. Pediatric aneurysms are different from adults in sex predilection, morphology, size, and location. Some pediatric aneurysms are giant (presenting at >25 mm) and are larger than many aneurysms in adults [6, 7]. Giant aneurysms are more frequently represented in the pediatric population [6, 8-10]. In childhood and adolescence, the rate of giant IA has been reported to be between 12% and 37% in the most recent case series [1, 6, 9-12]. Pediatric giant aneurysms are most often located in the posterior circulation and at the internal carotid artery (ICA) bifurcation, followed by the middle cerebral artery (MCA) bifurcation. The most common mode of presentation is subarachnoid hemorrhage (SAH) [13].

Based on a Pubmed search, we carried out a review of the literature from 2000 to 2017 regarding giant intracranial aneurysms diagnosed in the pediatric population. This descriptive study concerned patient number, sex, therapeutic management procedures and outcome (Table 1).

Our two patients presented with non-ruptured giant aneurysms misdiagnosed as carcinomas [14, 15] before operation. We discovered a couple reasons for the misdiagnoses. First, both of the patients were young women and they only presented with seizures or a persistent dull headache, no SAH. Based on their symptoms and the fact that adolescent aneurysm incidence is very low, we ignored the possibility of diagnosing an aneurysm before operation. Second, both of the two cases showed no SAH imaging in the CT or MRI examination. Case 1 showed a huge circular mass surrounded by an arachnoid cyst. An enhanced wall could be seen with an MRI enhancement scan so we considered the round mass and cystic cysts as a whole lesion, leading to misdiagnosis before operation. The MRI image of case 2 showed lesions located in the temporal lobe, and a T2 photo showed severe edema around the hematoma with no significant vascular flow-empty actions or tortuous vascular images. Combined with the huge lesions, we hypothesized it was most likely a glioblastoma with stroke. After the operation we reviewed the two cases and they showed something in common. The two cases both showed a clear boundary of the circular lesions in the MRI examination, and T1 and T2 photos both showed high signals which meant hematomas. Therefore giant aneurysms should have been highly suspected.

In case 2 the MRI image of the posterior high signal lesion was divided by a low-signal ring, which should be the thick wall of the aneurysm. There was no change in the low-signal ring in the contrast scan and most giant aneurysms rupture into the brain tissue. These two cases are extremely rare for young people, especially case 1. They remind us to consider a comprehensive diagnosis for such patients in the future, and that vascular examination is necessary to avoid misdiagnosis. This condition is rare and we should learn that aneurysmal SAH would not necessarily be included in the diagnostics of aneurysm when a child presents with seizure or headache.

The etiology of pediatric aneurysms remains unknown. Congenital structural changes of the vessel wall probably play a major role in aneurysm formation. Although the exact pathogenesis of aneurysm formation remains unclear, in our patient, no history of inflammation, traumatic events, connective tissue disorder, or polycystic kidney disease that can lead to aneurysm formation were found [5, 8, 16]. Krings et al [2] proposed that the pathogenesis of giant aneurysms differs from those of adult saccular aneurysms. The characteristic abrupt termination of the internal elastic lamina and muscular media at the entrance to adult saccular aneurysms at arterial bifurcations that is believed to be vulnerable to hemodynamic sheer stress and atherosclerotic or hypertensive degeneration was not found in an autopsy series of pediatric aneurysm specimens. Giant IA is a lesion that is regarded as a proliferative disease of the vessel wall with growth induced by extravascular activity. Recurrent bleeding from the vasa vasorum surrounding the giant aneurysm results in an increase in aneurysmal size and in
further proliferation of new membranes and vessels. This is in contrast to the previous view that intraluminal factors, such as change in hemodynamics and weak vessel walls, lead to the increase in size [17]. In summary, certain luminal and abluminal factors contributing to aneurysm formation were identified, but the pathophysiological mechanisms still need to be explored.

As with any other treatment of disease, the treatment of intracranial giant aneurysms has undergone major improvements over the years. Microsurgical and endovascular techniques are both available for the management of pediatric aneurysms. Many patients in this series received both endovascular and surgical therapies. Crossover between treatment modalities make drawing conclusions about the efficacy of either alone difficult. Currently, there are advocates for endovascular intervention in ruptured aneurysms in pediatric cases. The proficiency of endovascular intervention is increasing at a fast rate but long-term data are still lacking and the issue of coil durability remains a concern. Stiefe et al [18] in a case series of 13 pediatric patients over 12 years, showed that the results comparing surgical and endovascular intervention were equivocal. In 2012, Saraf et al [19] compared the treatment of pediatric aneurysms with open surgical treatment versus endovascular means. Morbidity and mortality were similar in both groups, but the endovascular treatment group had significantly higher rates of recurrence (14%) and formation of de novo (19%) aneurysms. Nik et al [20] believe the results of endovascular parent artery occlusion are not predictable and it has been shown that these aneurysms can continue to grow even after successful occlusion of the parent artery due to the persistence of vessel wall disease. Therefore they choose to treat aneurysms by surgical clipping.

Open surgical treatment of giant pediatric aneurysms present technical challenges. They are not amenable to direct clipping and advanced microsurgical skills are often necessary. Clip reconstruction of the aneurysm, aneurysm wrapping, and aneurysm trapping or excision with or without bypass have been described with good outcomes [8, 10, 21, 22]. Kalani et al [21] have described the Barrow Neurological Institute’s experience of 28 revascularizations performed for complex or giant pediatric aneurysms. They found good or excellent outcomes in 96% and complete aneurysm obliteration in 83% of their patients. An endovascular parent vessel occlusion approach may be suitable for a proximal giant aneurysm, and microsurgical management for the more distal cases using selective clipping with reconstruction, sometimes under protection of a cerebral revascularization. Because of the pseudo tumor syndrome and no SAH, we treated the aneurysms by surgical clipping followed by an excision. Both of the patients had excellent outcomes.

Giant MCA aneurysms in pediatric population are rare. Congenital defects in the vessel wall allow for the development of giant aneurysms, and they are prone to contain thrombi. In a CT or MRI scan they look like intracranial mass lesions and may be misdiagnosed as cranial carcinomas, so we have to account for the possibility of a giant aneurysm when there is a very clear and well-defined lesion in the inner brain tissue. Treatment of such aneurysms are challenging and may not be treatable by endovascular means. Here we report successful resection of two giant MCA aneurysms with anastomosis and reconstruction of the proximal and distal ends of the artery. Both of the patients had good outcomes. Therefore microsurgery is effective in the treatment of giant aneurysms in children.

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Disclosure of conflict of interest
None.

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References
[2] Krings T, Piske RL and Lasjaunias PL. Intracranial arterial aneurysm vasculopathies: target-
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[26] Krapf H, Schöning M, Petersen D, Küker W. Complete asymptomatic thrombosis and re-
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