Giant Serpentine Internal Carotid Artery Aneurysm: Endovascular Parent Artery Occlusion

A Pediatric Case Report

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Key words: giant serpentine aneurysm, cranial nerves dysfunction, 3D-XRA rotational angiography, parent artery occlusion

Summary

We report on a case of a 14-year-old boy with a giant serpentine aneurysm of the left internal carotid artery cavernous segment with symptoms of acute mass-effect cranial nerve dysfunction. After a balloon occlusion test of the collateral circulation, the patient underwent parent artery occlusion with platinum Guglielmi detachable coils and fibered coils. An optimal angiographic result and successful clinical outcome were achieved with resolution of IIIrd, IVth and VIth cranial nerve ischemic symptoms. CT angiography and 3D-XRA rotational angiography reconstructions gave sufficient inclusion information on the giant serpentine aneurysm angioarchitecture.

Introduction

Aneurysms are not congenital, not even in children. Aneurysms in the pediatric group are different from those in adults. There is a male dominance of 3:1, a higher incidence of unusual sites (posterior circulation 15%) and many aneurysms are large and giant in 20% with a lower morbidity rate. There is also a higher incidence of spontaneous thrombosis in the paediatric age group.

Giant aneurysms are about four times more common in children than in adults. In the paediatric age group, they are frequently associated

with other diseases such as aortic coarctation, polycystic kidney disease, fibromuscular dysplasia, Marfan syndrome, Ehlers-Danlos syndrome, etc.^{1,2}. Giant serpentine aneurysm was first defined in 1977 by Segal and McLaurin as partially thrombosed, giant aneurysms with an irregular vascular channel³.

Patient Characteristics

A 14-year-old boy, basic school student, without any history of neonatal or infant diseases was admitted for severe head pain accompanied by vomiting and abdominal pain to the surgical department for suspicion of appendicitis. Due to the severe head pain progression, vomiting, vertigo and rapid onset of IIIrd cranial nerve palsy, the patient was transferred to the Children's Neurological Department of the University Hospital. Initial CT scan confirmed a large mass lesion in the left cavernous parasellar region 30 mm in diameter with sphenoidal sinus and small wing defiguration. CT angiography suspected a large partially thrombosed cavernous aneurysm without any perifocal edema. Subarachnoid spaces were without signs of bleeding, the density of white and grey matter was also normal (figure 1). All biochemical analyses – biochemistry, coagulation, white and red blood cells count were normal.

Neurological examination confirmed complete IIIrd and IVth and incomplete VIth cranial nerve palsy with severe retrobulbar pain, prostration without hemiparesis. After neurosurgeon consults, DSA-angiography with a possible neurointerventional procedure was indicated.

Interventional Procedure

A 6F Terumo (Radiofocus, Tokyo, Japan) sheath was introduced into the right common femoral artery (CFA) and 5000 units of UF Heparin were administered intra-arterially. A 4F Vertebral Aqua – Tempo catheter was introduced over the Terumo .035/260 cm (Radiofocus, Tokyo, Japan) and selective carotid and vertebral angiograms were performed after that (figure 2).

Selective left common carotid angiography disclosed fibromuscular dysplasia of the left ICA cervical segment and a giant serpentine C4-C5 cavernous segment aneurysm was subsequently discovered distally. The C4 portion with an irregular thrombosed lumen was slightly elongated with a contrast agent intra-aneurysmal stagnation.

The left ophthalmic artery was filled at the thin C3 segment and distally, the left MCA territory was supplied with a clear delay without filling of the left ACA-anterior cerebral artery. The diameter of the left ICA giant aneurysm was 25x30mm, measured from the 3D-XRA rotational angiography. Transparent surface 3D-VR left ICA angiography was reconstructed, clearly showing the angioarchitecture of the cavernous giant serpentine aneurysm and its relationship with the left ophthalmic artery.

Selective right common carotid artery angiogram confirmed a normal cervical and intracranial picture of the right ICA with filling of the left ACA. Selective left VA-vertebral artery angiogram showed a supply of the left MCA territory through the large posterior communicating artery (PcomA).

Due to neurological symptoms of the progressive cranial nerves - III, IV, VI palsy and severe retro-ocular pain persistency a decision for left ICA occlusion test was proposed after discussions with neurosurgeons. A Corail 6F (Balt-Extrusion, France) guiding catheter with occlusion balloon at the distal tip was navigated into the left ICA. Another 4F Terumo (Radiofocus, Tokyo, Japan) sheath was introduced into the left CFA and supplemental 3000 units of UF Heparin were delivered. Left ICA occlusion test with 30 minutes of neurological status monitoring was achieved and additional collateral flow through the anterior and posterior communicating artery was displayed without manifestation of any additional neurological symptoms.

According to the good collateral flow confirmation, the left ICA parent artery occlusion (PAO) therapeutic treatment option was decided. A Surf 18 (William Cook Europe ApS, Bjaeverskov, Denmark) microcatheter was navigated into the giant serpentine aneurysm over Transend .014/300 (Boston Scientific corp.) and the embolization procedure was carried out. GDC 18-3D (Boston Scientific Corp.) four coils- 120 cms and DCS 18-spiral (William Cook Europe ApS, Bjaeverskov, Denmark), five coils - 110 cms were implanted directly into the aneurysm and the in-flow to the giant serpentine aneurysm with additional Tornado 18-8/3-14 coils was embolized. Final angiogram confirmed left ICA - C4 cavernous segment aneurysm occlusion, good collateral supply through the communicating arteries without any distal embolization and rapid filling of the left ophthalmic artery by the left PcomA (figure 3). Slow deflation of the guiding catheter tip balloon was done with continuous flushing throughout the following interventional procedures.

Patient follow-up and examinations

After the neurointerventional procedure, the patient was put on anticoagulation therapy with Clexane - (Enoxaparinum natrium) 0.6 ml s.c. one day dose, antiplatelet treatment with Plavix 75 mg per day, corticoids - with Dexona (dexamethasone) 8 mg i.v. application and analgesia continuous treatment. Slow verticalisation and physiotherapy was started 48-72 hours after the interventional procedure. Ophthalmological examination 14 days after the emboliza-

Figure 1 A-C) Native CT scan- hyperdense spheric lesion in the left parasellar region with sphenoid bone defiguration, partially thrombosed, 3cm in diameter. D,E) CTA in sagittal and coronal projections with partially thrombosed left ICA cavernous segment aneurysm. F) Complete III, IV and incomplete VI cranial nerve palsy with severe retroorbital pain.

tion confirmed a reduction in severe retrobulbar pain and a decrease in VIth cranial nerve palsy with improvement of left bulb lateral side movement. Cranial nerve palsy of the IIIrd and IVth nerves were unchanged. Electroencephalography was normal, neurosonological examination confirmed good left MCA, PCA, PcomA arterial flow and aneurysm thrombosis.

CT perfusion at 14 day follow-up demonstrated symmetrical hemispheric perfusion in CBF, CBV and MTT color maps (figure 4). At the six month follow-up improvement of IIIrd, IVth and VIth cranial nerve palsy with slight diplopia without retroocular pain was documented (figure 5). Ongoing left ocular physiotherapy training was proposed.

Figure 2 G,H) Left ICA DSA with fibromuscular dysplasia of the left cervical ICA and giant serpentine cavernous C4 segment aneurysm. Normal filling of the left ophthalmic and middle cerebral arteries. No filling of the A1 left ACA segment. I,J) 3D-XRA- rotational angiography – transparent reconstruction. K,L) AP of the left ICA and left lateral VA angiogram with PcomA collateral flow.

MRI and CE-MRA one year follow-up was performed with excellent collateral flow illustration predominantly through the left posterior communicating artery and less through the anterior communicating artery and A1 segment. MRI confirmed aneurysm sac thrombosis with aneurysmal diameter reduction without any flow. No perifocal edema or ischemia was seen (figure 6).

Discussion

In the largest, recently published paper by Lasjaunias et al¹, aneurysms in the pediatric population accounted for almost 6.8% of total aneurysms seen in all populations during the same screening period. They found a slightly higher male predominance 2:1 in patients over two years of age. In this group of 75 aneurysms in 59 pediatric patients, there was a total of 34 dissecting aneurysms (45%), 15 infectious aneurysms (8%), two chronic post-traumatic aneurysms in two patients (3%) and 24 saccular aneurysms (32%).

In the last few years the concept of aneurysmal vasculopathies has been strongly discussed with the influence on the therapeutic decisions

according to different etiologies of aneurysms and vascular wall diseases².

In 1977, Segal and McLaurin first introduced the concept of giant serpentine aneurysm, defined as partially thrombosed, giant aneurysms with an irregular vascular channel³. The natural history of GSA is still unclear, the majority of GSA did grow over time and hemorrhage occurred in 23.5%. Microscopic studies demonstrate that the aneurysmal wall consists of fibrous tissue measuring 2-4 mm in thickness often with calcification, loss or partial loss of internal elastic lamina and muscularis, with a number of small vessels. The sac contains thrombi of various ages with calcifications and haemorrhage.

The vascular intra-aneurysmal channels do not seem to be residual lumens of the parent artery but are typically intrathrombotic canals that are not endothelialized and do not contain normal elastic lamina or media. The canals may be central or eccentric, with typical slow flow^{4,5}. Clinical presentation of patients with GSA is of an intracranial mass effect rather than intracranial haemorrhage and is dependent on aneurysm location. In our patient, severe headache and cranial nerve palsy with subsequent oph-

thalmoplegia were also early signs of GSA due to ICA cavernous portion localisation. Rapid aneurysm growth, due to the vessel wall internal elastic lamina and muscular layer mistake may be suspected.

Therapeutic options for giant serpentine aneurysms depend on the aneurysm presentation, location and anatomic features of the feeding and draining vessels. The standard bal-

loon occlusion test provides information for the endovascular, surgical or combined approach. If good collateral flow or no vital distal branch artery supply occurs, the endovascular approach is easy. However, after endovascular occlusion of large aneurysms, symptoms of mass effect may persist^{6,7}. In our case the balloon occlusion test showed sufficient flow to the left MCA and ACA through the PcomA

Figure 3 M,N) 30 minute left ICA occlusion test with good cross AcomA and PcomA collateral flow without neurological symptoms before PAO treatment. O,P) Left ICA - PAO by coil embolization. Q,R) Final angiogram after PAO embolization procedure with good left ophthalmic artery and left MCA territory filling.

Figure 4 S) CTP-perfusion follow-up with symmetrical hemispheric perfusion in CBF, CBV and MTT color maps.

Figure 5 T) Six month follow-up with IIIrd, IVth and VIth cranial nerve palsy improvement with slight diplopia and without retroocular pain. Left ocular physiotherapy training ongoing.

Figure 6
One Year MRI and CE-MRA follow-up. U,V) Thrombosed left ICA aneurysm after PAO procedure without ischemia. W,X) CE-MRA of the Willis circle with the left MCA territory filling through the left PcomA and hypoplastic left A1-ACA.

and AcomA, so endovascular PAO therapy was available without the need for by-pass surgery. Symptoms of severe retrobulbar pain and cranial nerve dysfunction improved in the post-embolization follow-up⁸⁻¹¹.

Conclusions

The treatment of giant serpentine aneurysms should aim to arrest the growth of the aneurysm, eliminate the mass effect and obliterate

the abnormal vascular channel. The most effective way to accomplish this is direct and permanent occlusion of the parent artery at the origin of the aneurysm.

This can be achieved by selective catheterization of the artery and occlusion of the vessel with detachable coils, balloons or NBCA. If temporary occlusion of the parent artery is not tolerated, a surgical by-pass revascularisation procedure can be performed before permanent target artery occlusion.

References

- 1 Lasjaunias P, Wuppalapati S et Al: Intracranial aneurysms in children aged under 15 years: review of 59 consecutive children with 75 aneurysms. *Childs Nerv Syst* 21: 437-450, 2005.
- 2 Lasjaunias P, TerBrugge KG: Intracranial Aneurysms in Children. *Vascular Diseases in Neonates, Infants and Children*, Springer-Verlag Berlin Heidelberg: 373-392; 1997.
- 3 Segal HD, McLaurin RL: Giant serpentine aneurysms. Report of two cases. *J Neurosurg* 46: 115-120, 1977.
- 4 Anshun W, Feng L, Daming W: Giant Serpentine Aneurysms: Multidisciplinary Management. *Interventional Neuroradiology* 6: 41-52; 2000.
- 5 Nakatomi H, Segawa H et Al: Clinicopathological Study of Intracranial Fusiform and Dolichoectatic Aneurysms. Insight on the Mechanism of Growth. *Stroke* 31: 896-900, 2000.
- 6 Aletich VA, Debrun GM et Al: Giant Serpentine Aneurysms: A Review and Presentation of Five Cases. *Am J Neuroradiol* 16: 1061-1072, 1995.
- 7 Greene KA, Anson JA, Spetzler RF: Giant serpentine middle cerebral aneurysm treated by extracranial-intracranial bypass. Case report. *J Neurosurg* 78(6): 974-978, 1993.
- 8 Mawad ME, Klucznik RP: Giant Serpentine Aneurysms: Radiographic Features and Endovascular Treatment. *Am J Neuroradiol* 16: 1053-1060, 1995.
- 9 Zicherman J, Roychowdhury S et Al: Endovascular Treatment of a Ruptured Giant Serpentine Aneurysm of the Superior Cerebellar Artery in a Patient with a Chiari II Malformation. *Am J Neuroradiol* 25: 1077-1079, 2004.
- 10 Park JS, Lee MS et Al: Giant Serpentine Intracranial Aneurysms: A Base Report. *Korean J Radiol* 2(3), 2001.
- 11 Malisch TW, Guglielmi G et Al: Unruptured aneurysms presenting with mass effect symptoms: response to endovascular treatment with Guglielmi detachable coils. Part I. Symptoms of cranial nerve dysfunction. *J Neurosurg* 89: 956-961, 1998.
- 12 Mizutani T, Yoshimasa M et Al: Proposed Classification of Nonatherosclerotic Cerebral Fusiform and Dissecting Aneurysms. *Neurosurg* 45: 253-260, 1999.
- 13 Caramia F, Santoro A et Al: Cerebral Hemodynamics on MR Perfusion Images before and after Bypass Surgery in Patients with Giant Intracranial Aneurysms. *Am J Neuroradiol* 22: 1704-1710, 2001.

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EDITORIAL COMMENT

Prochazka et Al report on a rare disease in the pediatric population, i.e. a giant serpentine pediatric aneurysm presenting with symptoms of acute mass effect, and its successful endovascular treatment by parent vessel occlusion.

This case report emphasizes that although aneurysms are not congenital, in that they are not present at birth, they reflect structural or functional weaknesses of the vessel wall that are already present in utero and expressed at a later age, namely, in this case, fibromuscular dysplasia. This disease weakens the matrix of the blood vessel wall, and this intramural factor probably plays a very important role in the etiology of the reported aneurysm. Apart from intraluminal factors, other disease processes (or a combination of them) that may be responsible for the development of aneurysms can be found intramurally or on the outer vessel wall. In this regard, MRI in the acute stage would have been of particular interest since it may have shown signs of acute dissection or intramural hemorrhage further emphasizing the concept of chronic or acute dissections in the etiology of giant serpentine aneurysms.

In our opinion, giant serpentine aneurysm are subtypes of the partially thrombosed giant aneurysm type and are differentiated from them purely on morphological grounds. These aneurysms are characterized by perforators or cortical branches arising from the aneurysmal sac or an irregular ("serpentine"-like) vascular channel. They account for approximately 0.6% of all aneurysms and about 20% of all giant aneurysms. Their main clinical presentations are mass effect, ischemia and hemorrhage, although subarachnoid hemorrhage may occur in a quarter of cases. Giant serpentine or fusiform aneurysms occur typically in the adult age group and are more frequently encountered in males with no history of hypertension or atherosclerosis. The middle cerebral artery territory is most often affected followed by the internal carotid artery, the posterior cerebral artery and the basilar and vertebral arteries; other locations are exceptional. If not treated as described in the present case report, these aneurysms may grow over time or even rupture. Proximal occlusion of the parent vessel is the treatment of choice since it almost invariably causes complete thrombosis of the aneurysm. However, the possibility of a persistent dissecting process has to be kept in mind and close follow-up is recommended. Differential diagnoses of these aneurysms are dysplastic pseudoaneurysmal segments that are elongated into a dolichodysplastic segment, fusiform vessel ectasias seen after healed dissections, infectious aneurysms seen in HIV and fusiform aneurysms seen in connective tissue diseases. CT or MRI demonstrating thrombus in different ages and the patient's history should be sufficient to make these differentiations.

We conclude that congenital defects of the arterial wall combined with a dissection process as discussed above are the most likely underlying causes of giant serpentine aneurysm formation. These are exceedingly rare in the pediatric population and should be treated, as described in this report, by parent vessel occlusion and close follow-up to rule out persistent and progressive dissection.

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