Case Review

Cerebral aneurysms in childhood and adolescence

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Abstract: The authors present a series of eleven cerebral aneurysms in patients under the age of 18 years. All aneurysms were revealed by subarachnoid haemorrhage. These aneurysms can be distinguished from those occurring in adults with regard to their size, location and sex ratio inversion.

Eleven patients from 9-18 years of age, surgically treated, were included in this study. Patients' preoperative status was determined according to Hunt and Hess classification and Fisher grading (computed tomography scan). Cerebral angiography was routinely performed for diagnosis.

Two patients presented with multiple aneurysms. The male: female ratio was 1.75:1. All patients were in good preoperative grade (Hunt and Hess grades I to III). Only one aneurysm was located in the posterior circulation while 54.5% involved the internal carotid artery bifurcation. Mean aneurysm size was 10.5 mm.

Overall outcome was favourable in 8 patients (72.5%) without any sequelae and death occurred in 3 patients (27.3%). Causes of unfavourable outcome included vasospasm and brain swelling.

Aneurysms in this age group have specific characteristics concerning their size, location and sex ratio. Outcomes are similar to those in adults. Pathogenesis of cerebral aneurysms is also reviewed. *(p76-79)*

Key words: *Cerebral aneurysm, subarachnoid haemorrhage and paediatrics*

Introduction

Cerebral aneurysms in childhood and adolescence are rare neurosurgical lesions, occurring at a rate ranging from 0.5 - 4.6% of published aneurysm series.[5,14,17,19,21,23] Previous studies have described specific characteristics of aneurysms in this particular age group, including preferential location on the internal carotid artery (ICA) bifurcation, male predominance, and aneurysm's sac size. Clinical outcome is variable in the literature and some controversy still persists on the proportion of posterior circulation aneurysms.[8]

The aim of the present study was to focus on this specific age group and review our own surgical experience of 11 patients under the age of 18 years.

Material and methods

Eleven patients harbouring cerebral aneurysms were surgically treated in our institution between 1993-2003. Clinical presentation, radiographic findings and surgical management were assessed for each patient. Patients with subarachnoid haemorrhage (SAH) were classified according to Hunt and Hess grading.[9] Hunt and Hess grades I to III were considered a good preoperative status. Computed tomography (CT) findings were graded according to the Fisher classification. Each patient underwent a four-vessel angiography, using the Seldinger procedure with a femoral puncture. The three youngest patients of this series were explored by magnetic resonance angiography, but the lack of precision and definition of the actual devices led to an insufficient diagnosis.

All patients were admitted in a neurosurgical intensive care unit and were treated with a standard protocol including vasospasm prevention with nimodipine and systemic blood pressure management. No endovascular procedure was proposed as not available at the time of this study. All aneurysms were surgically treated by clip application within

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the first 72 hours following haemorrhage. The follow-up period ranged from 6 months to 10 years. The quality of clinical outcome was assessed using the Glasgow outcome scale (GOS).

Results

Thirteen cerebral aneurysms were treated in 11 patients under the age of 18 years.. The mean age at admission was 14.4 years (ranging from 9-18 years of age). There were 7 males and 4 females, resulting in a sex ratio of 1.75:1. This series represents 3.6% of cerebral aneurysms treated in our institution during the same period.

All patients of our series presented with typical subarachnoid haemorrhage. Preoperative clinical status was good in all patients, no grades IV or V of Hunt and Hess were noted: 63.6% were in grade I, 27.2% in grade II and one child in grade III. Only one patient suffered from seizures. Re-bleeding was noted in two cases (18.2%), occurring respectively 24-72 hours after the ictus, without modification of their clinical status.

On CT scans, 8 patients presented with grade II of Fisher (72.7%), one with grade III (9.1%) and 2 with grade IV (18.2%). Angiography led to the diagnosis of multiple aneurysms in 2 patients (two aneurysms in each case, including two mirror Sylvian aneurysms).

Distribution of aneurysms included 6 patients with ICA bifurcation aneurysms (54.5%), 2 aneurysms involved the anterior communicating artery (18.2%) and 3 were located on the major bifurcation of the middle cerebral artery (27.2%). Only one child had a basilar bifurcation aneurysm. Another patient treated for Sylvian aneurysm also had a posterior communicating artery aneurysm that disappeared on the postoperative angiography (probably thrombosed).

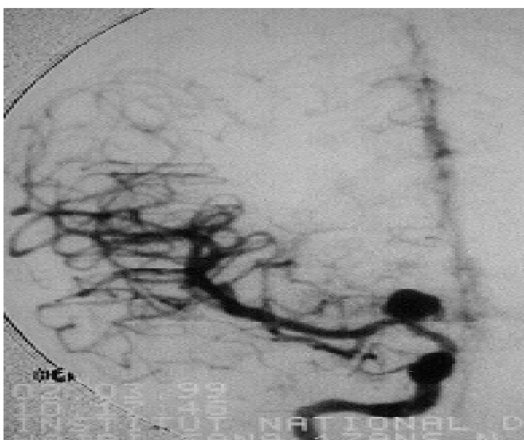
At admission, none of our patients presented with an unusual medical history or a history of head trauma. No mycotic aneurysms were observed, and we did not find any clinical signs suggesting autosomal dominant polycystic disease, neurofibromatosis, connective tissue disorder, aortic coarctation or haemoglobinopathy.

Aneurysm sac size ranged from 3 mm to 25 mm (mean size 10.5 mm). All children underwent microsurgical procedure including clip application via a pterional route, within the first three days after the ictus. Overall outcomes were favourable in 8 children (72.7%) with GOS score 5. Three patients died, 2 deaths were related to immediate postoperative brain swelling and the other one was due to severe vasospasm and diffuse cerebral infarctions (basilar trunk aneurysm).

Discussion

From a historical point of view, the first paediatric cerebral aneurysms description can be attributed to Eppinger in 1871, nearly one century after the first intracranial aneurysm report (Biumi in 1765).[4] MacDonald and Korb established the first paediatric series of cerebral aneurysm in 1939.[13] Since that report, about 725 cases have been published. Our study included 11 patients harbouring cerebral aneurysms under the age of 18 years, because their anatomical characteristics were different from their adult counterparts.[8]

Confirming the observations previously noted in the literature, the distribution of aneurysms seems to be particular to the paediatric age group. The ICA bifurcation (Figs. 1, 2 and 3) was the most involved location in our series (54.5%). This preferential location represents more than 25% of all aneurysms in a review of the literature. In Proust series of 22 children, ICA bifurcation was concerned in 36.4% of aneurysms, and according to this author, this is



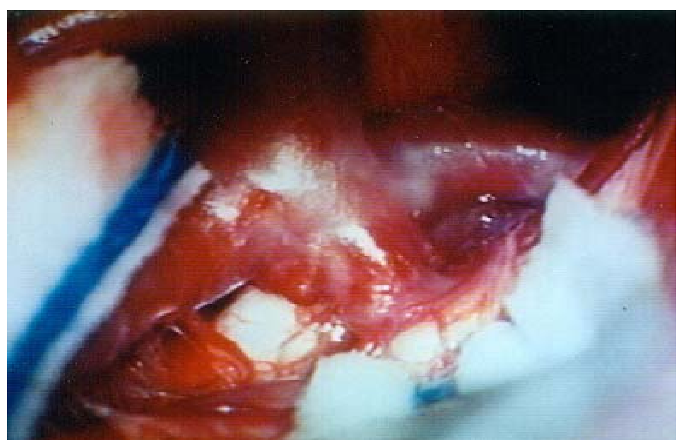


Figure 1 ↑ Frontal view angiogram demonstrating a saccular aneurysm of the right ICA bifurcation 54,5 % of our patients.
Figure 2 → Dissection and exposition of an ICA bifurcation aneurysm before surgical exclusion

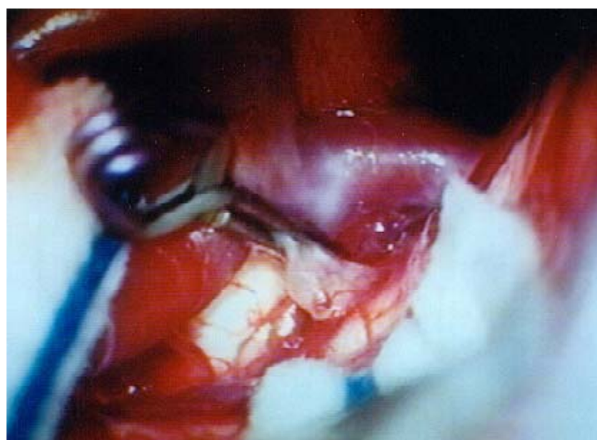


Figure 3 - Microsurgical exclusion of the ICA aneurysm with a Yasargil vascular clip and opening of the sac

the major topography of aneurysms in children with the exception of younger children.[20] This high frequency of ICA bifurcation aneurysms was observed in several studies: Storrs (31%), Pasqualin (29%), Robinson and Sedzimir (36%).[18,23,24]

The incidence of posterior circulation aneurysms remains controversial in children. Their frequency is high in some series by Amacher and Drake (46%), Storrs (35%) or Meyer (46%) while other studies have reported a proportion of 4-16% of vertebrobasilar aneurysms.[2,8,14,24] Frequency of giant aneurysm (superior to 25 mm) is also controversial. This latter reached 54% in Meyer series, 45% in Amacher and Drake study, and 31% in Storrs report.[2,24] Among our patients, only one giant aneurysm was observed. Overall aneurysm sac size seems to be more important in children than in adults.[8]

Re-bleeding was noted in 18.2% of our cases. Proust found 52% of aneurysms revealed by this complication.[9] Incidence of re-bleeding in the paediatric population is significantly higher than adults and is related to delayed diagnosis because of clinical polymorphism of SAH children.

Male predominance and inversion of sex ratio in paediatric aneurysm series is a well-known fact.[8] As observed in our patients, overall sex ratio is about 1.8:1 in all published series.

Pathogenesis of paediatric cerebral aneurysms remains hypothetic, since pathological studies concerning this age group are rare. It is classically known that vessel wall structural changes associated with haemodynamic stress lead to aneurysm occurrence. Congenital nature of these lesions has been noted by Lipper on the basis of large medial defect in the vessel walls observed in aneurysms occurring early in life.[11] Although we did not find this association in our series, some connective tissue diseases are well-known to initiate aneurysm development: Marfan syndrome, Ehler-Danlos type IV, neurofibromatosis type I, and autosomal dominant polycystic disease.[16] Infectious processes were also reported as potential causes of vessels parietal injury leading to less than 2% of cerebral aneurysms in children.[8] Finally, exceptional cases of post-traumatic aneurysms in children were described.[2,8] Their frequency seems to be more prominent than in adult population, reaching 14-39% of all paediatric aneurysms.[26] Post-traumatic aneurysms are preferentially located in the distal anterior cerebral artery. Finally aneurysms associated to aortic coarctation and sickle-cell anaemia remain exceptional.[3,19]

Treatment of paediatric cerebral aneurysms is similar to that of adults. New multimodal therapeutic strategies have led to a significant improvement of paediatric cerebral aneurysm management. In our study, 11 aneurysms were treated with surgical clip application, offering a 72.7% favourable outcome and a mortality rate of 27.3%. Management of paediatric aneurysms should include multidisciplinary staff, utilising endovascular techniques.[22,25] Surgical results are very heterogeneous in the literature (Table 1), and outcome seems related to preoperative clinical status as observed in adult series.[20] Cognitive and memory alterations (16%) could also occur, essentially after anterior communicating artery aneurysms treatment, as reported by Heiskanen and Vilkki.[6] Endovascular techniques have become an essential therapeutic approach, but the lack of long term follow-up in the published series remains a drawback in the assessment of this attractive method.[25] In the Sanai study comparing the durability of treatment following microsurgical and endovascular management of paediatric cerebral aneurysms, the authors found a comparable rate of morbidity

Table 1 - Review of recent large series of cerebral aneurysms in children and adolescents in the literature

Authors	Age limit	No. of cases	Good initial grade (%)	Posterior location (%)	Mortality (%)	Good outcome (%)
Amacher 1981	18	26	96	31	4	92
Heiskanen 1981	19	32	?	6.3	6	75
Storrs 1982	16	29	38	31	34.4	44.8
Ostergaard 1983	19	43	72	8	33	54
Roche 1988	16	43	81	16	12.2	79
Meyer 1989	18	24	50	46	4	92
Proust 2001	16	22	59	9.9	22.7	63.6
Present study	**18**	**11**	**100**	**9.1**	**27.3**	**72.7**

NB: Good initial grade includes I-III of Hunt & Hess classification. Good outcome means GOS 5.

and mortality in both groups.[22] They stated that microsurgery could be more effective in completely treating the paediatric aneurysm, compared to the 14% of recurrence in the endovascularly treated group after a mean follow-up of 5.7 years. Other authors noted a better outcome in patients treated by endovascular means.[1] We think that a multimodal approach should be discussed for each patient, and the surgeon should use the best treatment method he handles.

In our series, mortality was related to vasospasm and postoperative brain swelling, even if preoperative status in these cases was good. However, in the literature, amount of initial haemorrhage is the predominant cause of morbidity and mortality. Vasospasm appears to be better tolerated in this young age group, probably because of large collateral circulation functionality in younger patients.[8,20] This complication was observed in 53% of the 43 patients of Ostergaard, without any neurological deterioration.[17]

Conclusion

Cerebral aneurysms in young patients are distinct from their adult counterparts on the basis of sac distribution, aneurysm size and male predominance. Internal carotid artery bifurcation is the most found location in this age group. Polymorphism in SAH presentation in this age group leads to more delayed diagnosis than in adults. Discussion on the genesis of paediatric cerebral aneurysms is still hypothetic and requires further study of large pathological series.

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