INTRACRANIAL ARTERIAL ANEURYSMS IN CHILDHOOD

Case report

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ABSTRACT - A case of an intracranial arterial aneurysm at internal carotid bifurcation in a 10-year-old girl is described with the special features of cerebral aneurysm which occur in children, comparing with the adults. We alert for the necessity of carefully operative technique in order to avoid damage and intraoperative rupture of the aneurysm due to the very thin vessel wall that this population can develop. Our recommendation is early surgery in these patients.

KEY WORDS: intracranial aneurysm, pediatric cerebrovascular disease, subarachnoid hemorrhage, childhood.

Aneurismas arteriais intracranianos na infância: relato de caso

RESUMO - Relatamos o caso de aneurisma arterial intracraniano na bifurcação da carótida interna em menina de 10 anos de idade. As características especiais dos aneurismas intracranianos que acometem a faixa etária pediátrica são descritas, comparando com a faixa etária adulta. Alertamos a necessidade de emprego de técnica operatória microcirúrgica cautelosa para evitar lesão e ruptura intraoperatoriária do aneurisma, devido a parede do aneurisma geralmente ser muito fina na faixa etária pediátrica. Recomendamos cirurgia precoce nestes pacientes.

PALAVRAS-CHAVE: aneurisma intracraniano, doença cerebrovascular pediátrica, hemorragia subaracnóide, infância.

Intracranial aneurysms are rare in childhood, occurring at a rate of 0.17 to 4.6% in a population of patients who have undergone surgery for aneurysms1-4. The proportion of ruptured aneurysms in patients younger than 15 years is less than 1%5. In the pediatric population rupture of aneurysm is a cause of subarachnoid hemorrhage in 18-52.1%6,7, and several successful operations have been reported8.

In this report a case of a 10-year-old girl is described with the special features of cerebral aneurysm which occur in children, comparing with the adults.

CASE

A 10-year-old girl suddenly presented headache and a syncope episode while she was playing with dolls with her friends. Immediately she was left to hospital. At arrival she was GCS 13, isocoria, with stiff neck, without motor deficits (Hunt-Hess grade 3). At CT was subarachnoid hemorrhage (SAH) Fisher grade II and hydrocephalus (Fig 1). MRI showed aneurysm flow void (Fig 2) and arteriography showed a right internal carotid artery (ICA) bifurcation aneurysm with 4 mm (Fig 3). At the second day of SAH we performed a pterional approach and the aneurysm was clipped with a single straight clip. The wall of the aneurysm was very thin.

In order to control the hydrocephalus, we performed the opening of the lamina terminalis.

The outcome was excellent and 12 months later the patient is absolutely normal, with normal life activities.

Patient's mother gave written consent to publish her daughter case.

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DISCUSSION

Male preponderance in pediatric aneurysms has been reported in all large series as by Matson of 12:1, however most other authors have found a less dramatic distribution of 3:2, 5:2, 6:5. This finding is in contrast with adults whom a female predominance occurs. The female predominance in adult patients with aneurysm may be connected to the decreased level of estrogen in this group that results in a decrease of collagen, which plays an important role in maintaining the strength of the vessel wall.

Intracranial aneurysms are rare in childhood, occurring at a rate of 0.17 to 4.6% in a population of patients who have undergone surgery for aneurysms.
The incidence of aneurysms in children decreases with age: the proportion of ruptured aneurysms in patients younger than 20 years is approximately 3.5% and that in patients younger than 15 years is less than 1%\(^5\). Aneurysms in children younger than 5 years of age are rare and the majority of these cases occurred during the first 2 years of life\(^6,14,15\). The diagnosis of pediatric aneurysms at autopsy is exceptional\(^14\).

Rebleeding is frequently in the pediatric population occurring in a rate of 52-60%\(^14,17\). However, Ostergaard et al.\(^3\) showed a 13% rebleeding rate in his study of a pediatric population. Storrs et al.\(^15\), studied 29 children under age of 16 years and observed a syncope episode with meningeal signs in 62% of their patients, in half of these cases the rebleeding was the cause of neurological alteration. The incidence of rebleeding in the pediatric population is significantly higher than the 16 to 29% estimated in adult population\(^18\).

Most authors agree that the ICA bifurcation is the main location of aneurysms in children with rates of 24 to 50%\(^5,6,13,15\). In adults this location counts for a proportion of 2%. The main location on the circle of Willis is the segment distal to the first major bifurcation (M2) in about 40% of cases\(^2,3,6,15\). The frequencies of aneurysms in the posterior circulation and giant aneurysms remain controversial. Some authors\(^5,6,14,17\) have described a proportion of 0 to 16% of aneurysms in the verteobasilar system and others have reported 30 to 57%\(^6,19,20\). The prevalence of large aneurysms is about 50% compared with 27% in adults. In contrast, aneurysm in adults shows a predilection for proximal branches points and the anterior circulation. Multiple aneurysms are less common in children (3-5%) than in adults (10-20%)\(^21\).

The occurrence of aneurysms is probably the result of an interplay between congenital factors (changes in the vessel wall) and hemodynamic stress\(^21\). Lipper et al.\(^22\) suggested that a large congenital medial defect could be the initiating factor of aneurysms that occur early in life. Some inherited connective disorders have been associated with cerebral aneurysms: Marfan syndrome, Ehlers-Danlos syndrome, neurofibromatosis and autosomal dominant polycystic kidney disease. Crompton\(^23\) noted medial wall defects in cerebral arterial bifurcations that appeared to increase with age in his analysis of 149 autopsy cases. The degeneration starts in the intimal pads proximal to bifurcations, which then tended to extend to intima underlying the medial wall defects. It was stated that the intimal pathophysiological alteration by hemodynamic forces usually occurs at arterial bifurcations because is the site of the greatest shearing forces against the arterial wall\(^24\). It was demonstrated that large stress from the pulsating blood pressure produced structural fatigue in the arterial wall and degenerated the internal elastic lamina\(^25\). The role of hemodynamic factors is clearly illustrated by the preponderance of ICA bifurcation aneurysms is 24 to 50% and only 2.1% in adults\(^3,6,13\).

An infectious process and blunt trauma can produce a tear in the internal elastic lamina. Stehbens et al.\(^26\) suggested the fixation of bacteria at a site of traumatic aneurysms located in the area of tentorial incisure\(^27\). Previous reports have showed a higher incidence of mycotic and traumatic aneurysms compared to adults\(^28,29\). The incidence of mycotic aneurysm in the pediatric population is estimated to be 10%, whereas it is only 2.5% in the general population, frequently secondary to bacterial endocarditis. Three mechanisms of mycotic aneurysm formation have been postulated: local invasion of a vessel, infectious emboli to lumen of a vessel such as from bacterial endocarditis and cryptic mycotic aneurysm with no obvious source of infection. The most frequently isolated agents are *Staphylococcus aureus* and *Streptococcus*. A saccular aneurysm in a distal vessel or a fusiform aneurysm in a proximal vessel is suggestive of a mycotic aneurysm\(^29\). Mycotic aneurysms due to fungal disease tend to occur in immunocompromises patients and involve the large arteries of the brain. *Aspergillus*, *Candida* and *Phycomycetes* have been implicated\(^30\). Traumatic aneurysms are rare and are called pseudo aneurysms because they lack an endothelial lining. They tend to be supratentorial, peripheral, irregular in contour and do not have necks. Of the traumatic aneurysms, 75% occur in children less than 16 year. Intracranial vascular anomalies (fenestrations, duplications and accessory vessels) and intracranial aneurysms are associated\(^31\). Based on these facts, the development of a cerebral aneurysm in children is considered to be an association between an acquired factor, hemodynamic stress and congenital defects of the vessel wall.

In adults, headache is the most common presenting symptom. Infants present with irritability, lethargy, vomiting, seizures and coma. Children present with symptom due to aneurysmal rupture as opposed to mass effect on the brain from the aneurysm. Cerebral aneurysms in childhood have been associated...
with head trauma (including birth trauma), infection (mycotic aneurysms), autosomal dominant polycystic kidney disease, fibromuscular dysplasia, subacute bacterial endocarditis, coarctation of the aorta, collagen vascular disease, Ehlers-Danlos syndrome, Marfan syndrome, syphilis, sickle cell anemia and tuberous sclerosis. These associated conditions are commonly found in children with symptomatic aneurysms.

Allison et al. made a retrospective review about aneurysms in a pediatric population and their correlations with imaging. They showed that conventional angiography remains the gold standard for preoperative imaging, but CT, MRI, and MR angiography (MRA) frequently reveal aneurysms. CT may show focal hyper dense mass with or without enhancement in addition to hemorrhage. MRI may show a focal mass with different signal than the surrounding hemmorhage with or without flow void or enhancement. MRA can define aneurysms as small as 3-4 mm. The limitations of MRA included no visualization of an aneurysm secondary to adjacent spasm, small aneurysm size and presumed slow flow in the aneurysm. A major role of MRA in the future may be in the noninvasive screening evaluation of patients at high risk for developing aneurysms. If surgery is not performed immediately, MRI and MRA are important to noninvasive follow the aneurysms.

Obliteration of the aneurysmal sac with a metal clip is the definitive treatment of cerebral aneurysms in any age. However, because of the small size and fragility of children vessel other techniques can be used like microanastomosis, bypass procedures, hypothemic arrest for giant aneurysms, the endovascular approach and the basilar artery occlusion. With regards to neonates and infants, endovascular treatment of children with vein of Galen malformations or AVMs is generally accepted.

Morbidity and mortality is lower in children presumably due to less vasospasm and underlying atheroesclerotic disease. The mortality after the first cerebral hemorrhage in the first 48h ranges from 14 to 34% in adults compared with 11-12% in early childhood. An intracerebral hematoma occurred in 37% of children, complicating 42% of MCA aneurysms. Vasospasm is the second cause of mortality after SAH and occurs in a rate of 53% (3) and is well accepted in the most part of children.

The percentage of good outcome varies between 63.5 to 75%. However the rate of excellent results rises in children with good preoperative grades to 85 to 95%. These very good outcomes obtained and the high risk of rebleeding in children with cerebral aneurysm allows the recommendation of early surgery in these patients.

Our pediatric patient had presented an unusual intracranial pathology for her age: an aneurysm. Its topography (internal carotid artery bifurcation) is common and the very thin aneurysm domus wall has been described in infancy. The good outcome is described too.

We alert for the necessity of carefully operative technique in order to avoid damage and intraoperative rupture of the aneurysm due to the very thin vessel wall that this population can develop and the recommendation of early surgery in these patients.

REFERENCES


