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Vanishing of A Congenital Intracranial Aneurysm

Intracranial aneurysms in neonatal period and early infancy may be resorbed spontaneously or due to removal of predisposing factors. Herein, we presented a rare case of vanishing of a large thrombosed intracranial aneurysm in a 25-day-old girl presented with head enlargement and vomiting. Magnetic resonance imaging (MRI) revealed a severe hydrocephalus accompanied by a 1.5-cm aneurysm at the bifurcation of the right middle cerebral artery (MCA). Doppler ultrasonography confirmed a flow with a magnitude almost similar to that of the right MCA flow, inside the mass located adjacent to it. The hydrocephalus was managed by ventriculo-peritoneal (VP) shunt. The patient underwent a conservative observation for several months. Subsequently, the aneurysm was vanished in control images.

Keywords: intracranial aneurysm, hydrocephalus, congenital, thrombosis

Introduction

Intracranial aneurysm is an extremely rare condition in childhood, especially in early infancy and neonatal period.[1,2] It has been estimated that only 0.005%–2% of all diagnosed aneurysms occur in pediatric age group.[3] However, this uncommon condition may lead to disastrous events such as subarachnoid hemorrhage (SAH), intracranial hemorrhage (ICH), subsequent brain damage, and death.[4] A high proportion of patients with intracranial aneurysms demonstrate neuroradiologic features of complete or partial spontaneous intra-aneurysmal thrombosis. Such phenomenon is probable to be of little significance causing no alteration in outcome[5], although in occasionally, it might be associated clinical symptoms.

There are only a few reports of vanishing of intracranial aneurysms following complete thrombosis of the lesions.[6,7]

Herein, we reported on the youngest case of spontaneous resorption of a congenital large intracranial aneurysm that developed asymptomatic thrombosis and was vanished within several months.

Case Presentation

A 25-day-old girl, the product of an uneventful 38-week gestation from healthy and nonrelative parents, was referred to Children's Hospital Medical Center with progressive head enlargement and occasional vomiting. The pregnancy had been totally normal, and the child was born by cesarean section due to enlarged head circumference (HC) of 38 cm at birth. The postnatal period was accompanied with increasing HC by more than one cm per week.

Initial clinical examinations revealed an otherwise healthy infant with bulge and wide anterior fontanel, sunset eyes, widening of cranial sutures and HC of 42 cm, but normal neurologic findings.

A brain magnetic resonance imaging (MRI) which had been performed at the age of 14 days to evaluate the hydrocephalus, revealed severe three-ventricular

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hydrocephalus due to aqueductal stenosis, and very thin brain cortex, with the right lateral ventricle being more dilated than the left one. Additionally, the hydrocephalus was accompanied by a 1.5-cm signal void mass appearing at the bifurcation of the right middle cerebral artery (MCA) with no evidence of hemorrhage. In T1-weighted image, the mass lesion was heterogeneous with isointense core and hyperintense periphery, suggesting the pattern of a low-speed blood flow through the lesion and peripheral thrombosis (Fig. 1). Proton density image demonstrated a 1.5 cm very hypointense lesion at MCA bifurcation with posterior and slightly superior projection (Fig.2). Brain magnetic resonance angiography (MRA) was not performed at the time of the first MRI and was postponed till the patient's general condition would improve and she could tolerate the long time and anesthesia necessary for MRA. For lack of facilities to perform the standard angiography for neonates and young infants in our setting, a transcranial Doppler ultrasonography was performed on admission, which confirmed a flow with a magnitude almost similar to that of the right MCA flow, inside the mass located adjacent to it (Fig. 3).

The hydrocephalus was managed with standard ventriculo-peritoneal (VP) shunt. Intracranial pressure was considerably high during ventricular catheter insertion. The cerebrospinal fluid (CSF) parameters including analysis and culture were normal. The post-operation period was uneventful. Subsequently, her clinical signs and symptoms were subsided and the HC was decreased.

The follow-up cranial sonography performed three months later revealed an increased cortical thickness and no flow in the MCA aneurysm. Since there was no evidence of abnormal flow in the same area and the patient's symptoms were recovering in a good way, we preferred to approach the case more conservatively and postponed angiography to an older age. MRA performed six months and one year after the operation demonstrated no abnormalities in the right MCA territory (Fig. 4). Meanwhile, a control MRI confirmed total resorption of the lesion at the same levels (Fig. 5). Due to normal findings in MRA and MRI at age of one year and for lack of good facilities for four-vessel digital subtraction angiography (DSA) for young patients in our center, the parents did not

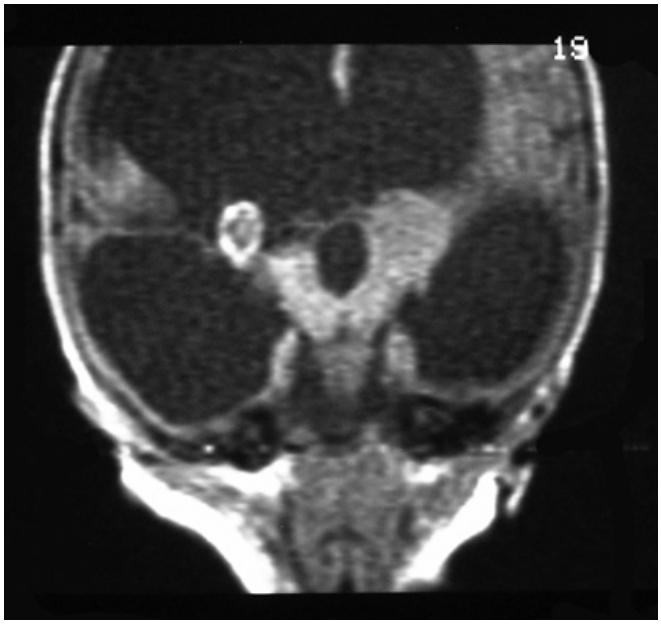

Fig. 1. The coronal view of T1-weighted image from the pre-operative brain MRI confirms a heterogeneous mass on the place compatible with the right MCA bifurcation.

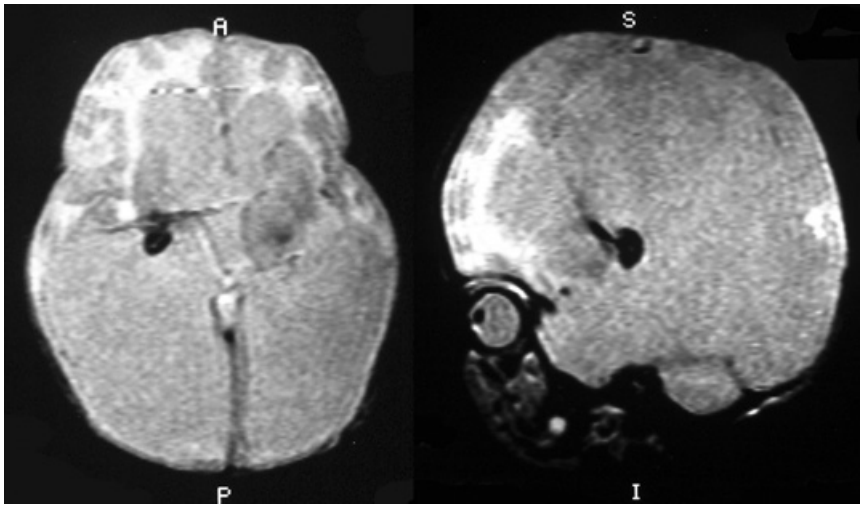

Fig. 2. A and B. Axial and sagital views of proton density images revealed a signal voiding mass on the bifurcation of the right MCA confirming a large aneurysm.

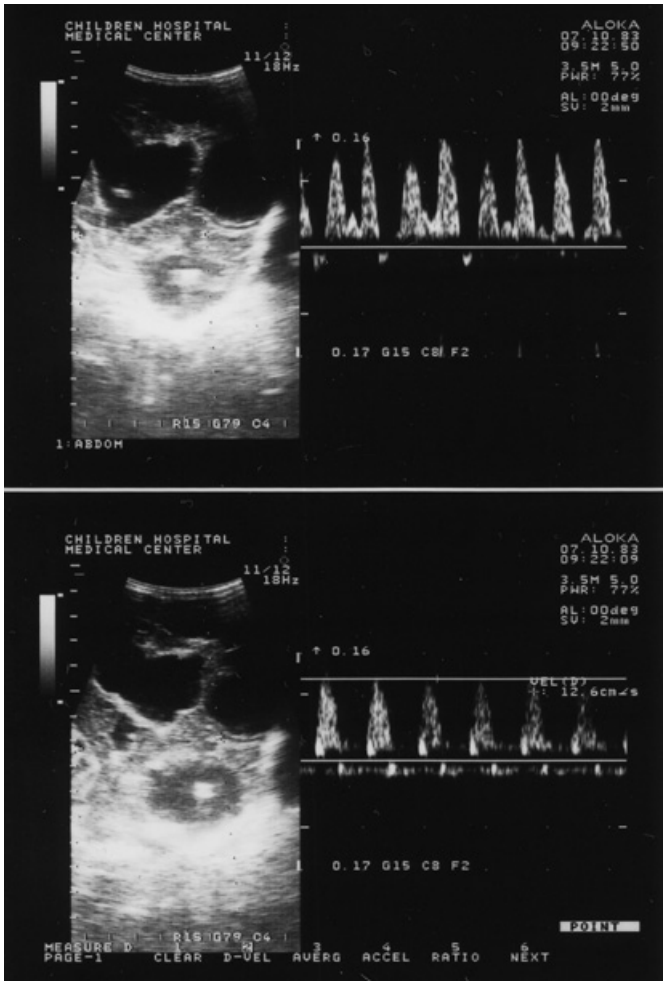

Fig. 3. Doppler ultrasonography on admission confirmed a flow with a magnitude almost similar to that of the right MCA flow, inside the mass located adjacent to it.

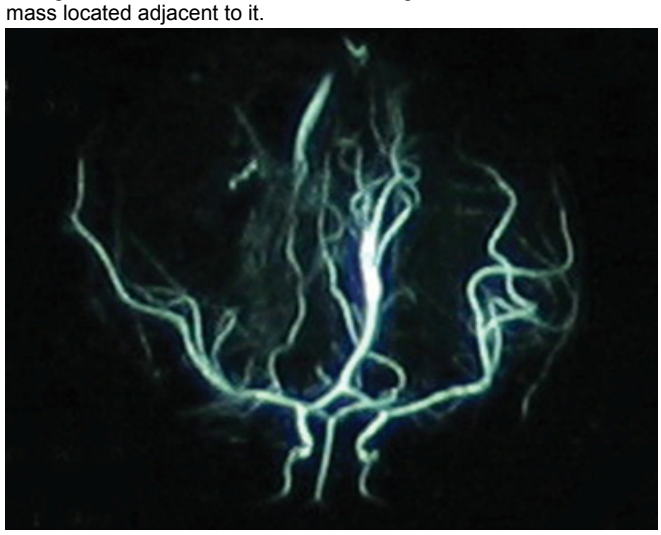

Fig. 4. Brain MRA at the age of one year shows normal vasculature, especially in the right MCA territory.

accept the probable complications, and refused to perform DSA.

Now, the girl is two years old with an HC of 49 cm. She is completely healthy, can sit and walk alone, and talk several words, with the neurologic findings being totally normal.

Discussion

Pediatric intracranial aneurysm is different from that of adults in several ways such as sex ratio, size, location and etiology. In contrary to adults, there is a male predominance in children with a male:female ratio of 4:3.[8] Giant aneurysms—*i.e.*, those >2.5cm in diameter—have been documented to be more common in children presented more in association with seizures and mass effect. Moreover, mycotic and traumatic aneurysms account for a much higher proportion of childhood aneurysms than do those that present in adulthood[7]. Furthermore, pediatric intracranial aneurysms tend to occur more in internal carotid, MCA and anterior communicating arteries.[9]

Clinical presentations consist of vomiting, seizures, hydrocephalus, mass effect, cerebrovascular accident, SAH, and coma. SAH is still the leading clinical manifestation of cerebral aneurysms.[8] Even though, the incidence of aneurismal SAH has been shown to be less than 1% in the first decade of life.[10]

The issue of whether pediatric aneurysms are congenital or acquired has been the matter of conflict for many years. Some authors assume them to be congenital while others consider them an acquired degenerative condition.[8,11-13]

The inclination of large aneurysms to develop partial or total thrombosis has been well described in the literature.[14] Although partial thrombosis is a common event in giant aneurysms accounting for 48% to 76% of patients, complete thrombosis is extremely rare.[7] There are some potential theories regarding the causes of thrombosis in cerebral aneurysms, the most important of which is likely the relationship between the volume and neck size of the aneurysm; a small orifice and a large volume result in slow flow within the lumen and spontaneous clotting.[15] The contrast medium used for angiography may cause thrombosis, probably due to intermittent vasospasm or dehydration caused by its hyperosmolarity.[7] The endothelial damage induced by turbulent blood flow within the sac and subsequent intramural hemorrhage may bear a crucial role in the development of thrombosis in giant aneurysms. Aggregation of platelets to the subendothelial matrix can accelerate formation of

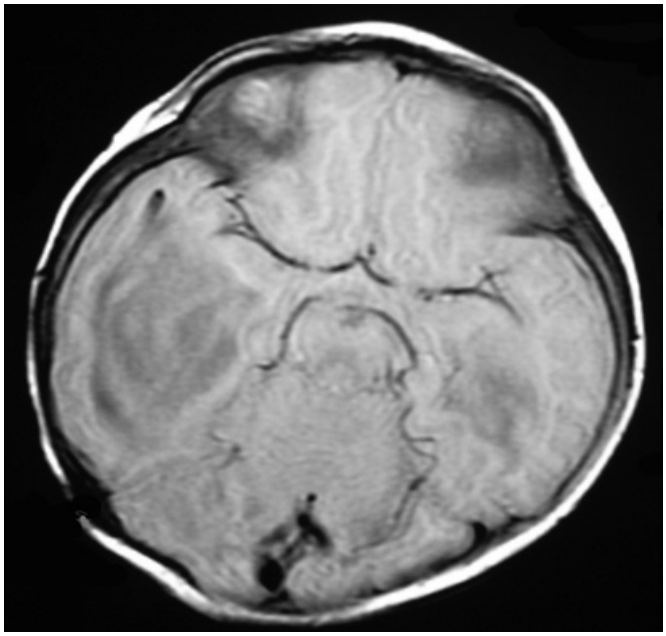

Fig. 5. Proton density image of patient's brain MRI demonstrates normal right MCA and its distribution. This image has been provided at the same level of axial view of Figure 2.

thrombus.[16]

Complete thrombosis of a giant aneurysm is an ongoing dynamic process, which has potential for further growth and producing mass effect due to accumulation of thrombotic materials, recurrent intramural hemorrhage, or development of intrathrombotic capillary channels.[17-20]

It is well-documented that either thrombosed or non-thrombosed giant aneurysms should be managed regardless of the presence of intraluminal clot.[5] On the other hand, there is one report of vanishing of a giant intracranial aneurysm in a 9-month-old girl following its complete thrombosis.[7]

In this case report, a young infant with unruptured aneurysm and severe hydrocephalus has been presented. We have tried to go through the pathogenesis of the condition and categorize the possible hypotheses:

-Although in the majority of such cases, hydrocephalus is secondary to SAH, the fact that the aneurysm was unruptured potentially proved that the hydrocephalus was a primary congenital condition. This argument can confirm that the aneurysm was an associated congenital problem due to a structural defect in intracerebral development.

Considering that the embryonic development of cerebral vessels might be influenced by hemodynamic factors, it would be postulated that in our patient with dilated right temporal and frontal horns, the blood flow in the MCA while passing through the Sylvian fissure developed some disturbances in the MCA trunk or its bifurcation resulting in vessel wall damage and aneurysm formation. Moreover, after managing hydrocephalus by VP shunt, as a result of establishment of normal intracranial pressure in the right ventricle, the vessel wall obtained the opportunity to return to a normal state and the aneurysm was regressed and vanished spontaneously.

-Assuming that there had been probable vascular defects such as fragmentation of the internal elastic membrane or the muscular layer of media at MCA bifurcation,[14] it is possible that the premature structures of vessels have developed gradually during the first months of life as well as strengthening the weak segments of cerebral vessels through the maturation process. This process has probably resulted in aneurysm resorption.

It can be presumed from the case that, although intracranial aneurysm is a risky condition, in very young children, it is possible that it resorbs spontaneously either following thrombosis or like this case, after correction of the predisposing factor. Therefore, if there are not enough facilities to do angiography or operation while the patient's images show the evidence of thrombosis, it may be feasible to postpone aggressive management and follow the patient to older ages. It may give the lesion enough time to regress naturally or for removing the causative factors.

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