Intracranial Aneurysms Associated with Moyamoya Disease in Children: Clinical Features and Long-Term Surgical Outcome

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■ BACKGROUND: **Moyamoya disease (MMD) in children was rarely associated with intracranial aneurysms. The purpose of this study was to report the clinical characteristics and long-term surgical outcomes of pediatric intracranial aneurysms accompanied with MMD.**

■ METHODS: **Between October 2002 and October 2013, our department treated 9 pediatric MMD patients (aged ≤17 years) with intracranial aneurysms. Clinical and angiographic features, treatment selection, as well as follow-up information were obtained and analyzed. The efficacy of vascularization and the changes of intracranial aneurysms were evaluated with digital subtraction angiography (DSA). We also collected 7 previously published reports to analyze the characteristics of this rare condition.**

■ RESULTS: **In our series of 9 patients, 7 were male. The mean age was 11 ± 3.4 years (range 5–16). Seven patients presented with intracranial hemorrhage as the initial manifestation, while 2 patients suffered transient ischemic attacks. The most common aneurysm location was the posterior choroidal artery (4, 44.4%). One anterior choroidal artery aneurysm was completely embolized with Onyx (ev3, Irvine, California, USA). One posterior choroidal artery aneurysm failed due to inaccessibility to the parent artery. Bilateral encephalo-duro-arterio-synangiosis (EDAS) surgery was performed for all the children. During the follow-up period of 6.4 ± 2.2 years (range 3–11), spontaneous occlusion of aneurysm was observed in 4 children, including 1 child with middle cerebral artery aneurysm, 1 with lenticulostriate artery aneurysm, and 2 with posterior choroidal artery aneurysm. Good or fair vascularization was observed in all the 9 children with DSA follow-up. No patients suffered intracranial hemorrhage during the follow-up period.**

■ CONCLUSIONS: **The long-term survey showed EDAS surgery could effectively increase the cerebral blood flow and maintain good outcomes in children, which may further result in the disappearance of the intracranial aneurysms and decrease the incidence of recurrent hemorrhage.**

INTRODUCTION

The incidence of intracranial aneurysms in moyamoya disease (MMD) has been estimated to be 3%–14% in adults.[1] A large number of cases in the adult population have been reported, whereas the disease is uncommon in children. To our knowledge, aneurysm associated with MMD can be classified into 2 types: the major artery aneurysm (60%) and blood flow secondary to the arterial stenosis (40%; choroidal, moyamoya vessels, meningeal vessels).[2]

Aneurysms on the major artery aneurysm usually present with subarachnoid hemorrhage, while aneurysms on the peripheral arteries are associated with intracranial or intraventricular hemorrhage.[2-6] Additionally, a minor proportion of children present with an incidental finding or manifest with symptoms of transient ischemic strokes.

During the past decades, the natural history and treatment selection of these aneurysms in pediatric MMD patients have been rarely reported and remain controversial.[7-12] Our former article reported the abnormal dilation of anterior choroidal artery, and

Key words
- Aneurysm
- Intracranial
- Long-term
- Pediatric
- Surgical outcome

Abbreviations and Acronyms
DSA: Digital subtraction angiography
EDAS: Encephalo-duro-arterio-synangiosis
MMD: Moyamoya disease

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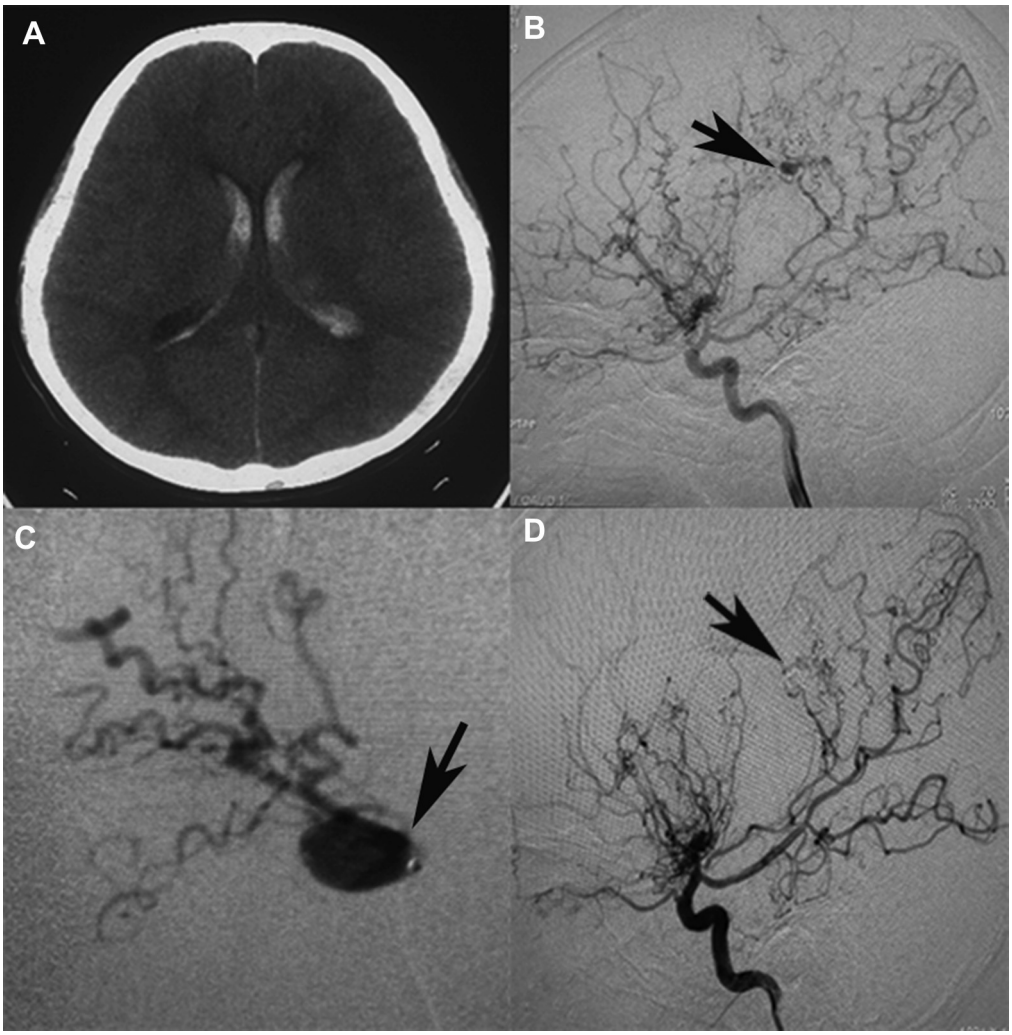

Figure 1. An 11-year-old boy with moyamoya disease (MMD) suffered a sudden onset of intracranial ventricular hemorrhage with predominance of hematoma in the left ventricle (**A**). The lateral angiogram of the left internal carotid artery showed the presence of an anterior choroidal artery aneurysm (*arrow*) (**B**). The superselective angiogram through a Marathon microcatheter showed the aneurysm was associated with a pial vessel (*arrow*) (**C**). The postprocedural lateral angiogram of the left internal carotid artery showed the complete occlusion of aneurysm sac (*arrow*) (**D**).

posterior communicating artery may be the major cause of hemorrhage in children absent of intracranial aneurysms. However, children with the intracranial aneurysms were not included in the study.[13] Previous data showed that blood flow modification after revascularization could lead to spontaneous regression and disappearance of these aneurysms.[3,14,15]

The purpose of this study was to report the characteristics and long-term follow-up outcomes of pediatric intracranial aneurysms with MMD.

MATERIALS AND METHODS

Study Population

The study protocol was approved by the research ethics committee at Beijing Tiantan Hospital and 307 Hospital PLA. Formal written consent was obtained from all patients. Between October 2002 and October 2013, 469 pediatric MMD patients were treated in our hospital. Among them, 9 (2 females and 7 males) had intracranial aneurysms with an incidence of 1.9% (9/469). Clinical, radiographic features and the treatment selection were retrospectively reviewed for each patient. The diagnosis of MMD met the current criteria. Intracranial hemorrhage was confirmed with computed tomography and cerebral infarction with magnetic resonance imaging (MRI).

Endovascular Treatment

Two children underwent endovascular treatment under general anesthesia after giving informed consent. Both procedures consisted of transarterial embolization using Onyx (ev3, Irvine,