CLINICAL ARTICLE

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Encephalo-duro-arterio-synangiosis(EDAS) using Occipital Artery in Children with Moyamoya Disease

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Objective : Although an encephaloduroarteriosynangiosis procedure using the superficial temporal artery (STA-EDAS) is an effective indirect bypass method in children with moyamoya disease(MMD), there is still a need for an additional bypass operation that can cover the area of the posterior circulation. The goal of this study is to evaluate the efficacy of the EDAS procedure using the occipital arteries (OA-EDAS).

Methods : From August 2003 to April 2004, We performed OA-EDAS in sixteen patients with MMD who have a circulatory insufficiency in the territory of the posterior cerebral artery(PCA). The medical records were reviewed retrospectively. The surgical outcomes, including the changes in neurological status and imaging studies, with the degree of neovascularization on the cerebral angiogram, and the hemodynamic changes on single-photon emission computed tomography(SPECT), were analyzed.

Results : These 16 children consisted of 5 boys and 11 girls aged 2 to 9 years. The clinical outcome of their PCA symptoms, such as visual transient ischemic attacks(TIAs) or visual field defect, was favorable in 14 patients of 16. Nine patients of 11 who underwent follow up magnetic resonance imaging(MRI) showed favorable MRI changes. On angiogram most of the patients exhibited good or fair revascularization of the PCA territory (7 of 8). The hemodynamic changes on SPECT in the PCA territory after surgery showed improved vascular reserve in 13 of the 16 territories.

Conclusion : OA-EDAS is a safe and efficacious revascularization procedure in patients with MMD who have compromised cerebral perfusion in PCA territory, or with visual TIAs.

KEY WORDS : Children · Moyamoya disease · Occipital lobe · Revascularization.

Introduction

Moyamoya disease(MMD), the most common pediatric cerebrovascular disease in eastern Asia, is characterized by progressive occlusion of the bifurcations of internal carotid artery and posterior cerebral arteries, accompanied by the formation of extensive collateral vessels at the base of the brain. The clinical presentation of MMD usually includes repeated TIAs in children and intracranial hemorrhage in adults[2,20]. Although no curative treatment is available (because of its unclear pathogenesis), the benefits of revascularization surgery for the ischemic type of MMD are well established[2,5,8,17].

The goal of bypass surgery is to establish adequate collateral circulation to ischemic brain tissue. Most surgical approaches have focused on increasing the blood supply primarily in the middle cerebral artery(MCA) and anterior cerebral artery (ACA) territories[1,7,12,14,17]. However, these approaches do not directly benefit the PCA territory.

Revascularization of the PCA territory should be considered for the treatment of pediatric MMD, for several reasons. First, it is well known, on the basis of various clinical and angiographic findings, that the disease process in children is dynamic and progressive[20], and this progression eventually involves the PCA[1,6,21] ; therefore, the deterioration of the blood flow in the PCA territory may continue to progress, even if there is good collateral formation in the MCA territory. Second, impaired circulation in the PCA territory is relatively frequent. Moreover, in a report by Kim et al., visual symptoms were seen more often in patients with a juvenile onset than in cases of adult onset[11]. In a report by Suzuki et al. although no infarcted foci of the occipital lobe were observed on the computed tomographic(CT) or MRI scans, many of the patients with MMD

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showed evidence of significantly decreased metabolism and CBF in the PCA territory, as revealed by their positron emission tomographic and acetazolamide SPECT scans[21]. Third, the occipital area is essential for visual function. Ischemic brain damage in the PCA territory may lead to visual disorders, directly affecting the patient's quality of life[5,6]. Finally, bypass surgery to treat childhood MMD should be performed before irreversible ischemic damage occurs, because it is known that no development of collateral vessels occurs after bypass surgery in already infracted areas[19].

These multiple infarctions represent the main reason for poor clinical outcomes[8]. This situation is the same in the PCA territory. Therefore, prophylactic revascularization surgery should also be considered for the PCA territory. We performed EDAS using the occipital artery (OA-EDAS) for Moyamoya patients with PCA involvement, and analyzed the results in terms of their clinical outcomes, neuroimaging changes, the extent of revascularization on the angiograms, and the hemodynamic changes on the SPECT scans.

Materials and Methods

We treated forty-nine children with MMD between Aug. 2003 and April 2004. Of these, sixteen children were treated with OA-EDAS. We performed OA-EDAS in MMD patients with visual symptoms (e.g. visual cognitive dysfunction symptoms, visual field defects, attacks of transient blindness and visual hallucinations) with PCA stenosis and/or a decrease of vascular reserve in PCA territory on the SPECT scan.

The diagnoses were preoperatively confirmed for all of the children with the use of MRI and all patients underwent a conventional angiographic evaluation for the study of the intracranial and extracranial carotid arterial supply and confirmation of the diagnosis of MMD. The preoperative angiographic stages were evaluated according to the classification described by Suzuki and Kodama[20]. All 16 patients underwent preoperative SPECT scanning with 99mTc-HMPAO and acetazolamide SPECT scanning. All 16 patients underwent OA-EDAS in conjunction with STA-EDAS and bifrontal encephalo-galeo-(periosteo)-synagiosis (EG(P)S). The follow-up periods ranged from 4 to 18 months (mean 11.8mo.)

The clinical improvements in the PCA territory were classified into two groups. Patients in the favorable groups exhibited complete recovery of the PCA ischemic symptoms or preservation of normal visual function, whereas those in the unfavorable group exhibited persistent or worsened symptoms. Eleven patients underwent follow-up MRI assessments, 5 to 17 months (average, 10.1mo.) after the last operation. The postoperative MRI findings were compared with the preoperative findings and classified into two categories. In the favorable category, the MRI demonstrated no infarctions in either the pre- or post operative scans or no further increase in the infarction size. In the unfavorable category, neuroimaging demonstrated an increase in the infarction size or the formation of a new lesion.

Eight patients underwent follow-up angiograms, 3 to 10 months (average, 5.4mo.) after surgery. The development of collateral circulation in the PCA territory (n=8) through the OA-EDAS was graded according to the system described by Matsushima et al.[13] ; a good score represented revascularization of more than two-thirds of the PCA distribution, a fair score represented revascularization of one- to two-thirds of the PCA distribution, and a poor score represented slight or no revascularization (Fig. 1).

All sixteen patients were evaluated for hemodynamic changes. The average interval between the operation and the last SPECT examination was 10.9 months (range, 4 to 18mo.). The postoperative SPECT findings for the PCA territory were compared with the preoperative findings and classified into one of two groups. The favorable group included patients for whom SPECT scans demonstrated no hemodynamic abnormalities either pre- or postoperatively or a decrease in the size of hemodynamic abnormalities. The unfavorable group included patients for whom the SPECT scans demonstrated no change in the size of hemodynamic abnormalities or revealed new hemodynamic abnormalities. The hemodynamic abnormalities taken into consideration included perfusion defects, decreased perfusion, and decreased vascular reactivity of the cerebral vessels to acetazolamide.

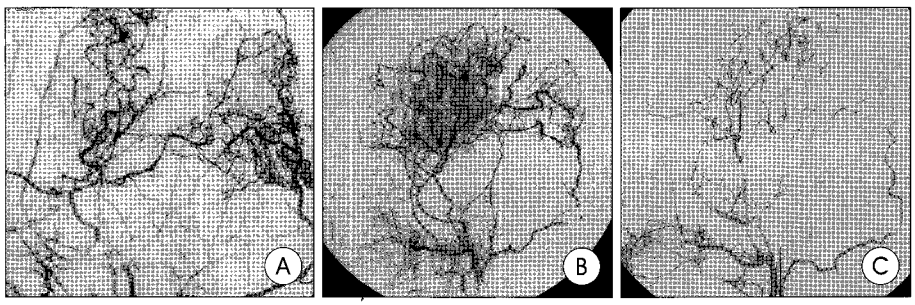

Fig. 1. Angiogaphic classification of the development of collateral circulation in the posterior cerebral artery (PCA) territory through the bypass. A : Good, with revascularization of more than two-third of the PCA distribution. B : Fair. with revascularization of one- to two third of the PCA distribution. C : Poor, with slight or no revascularization.

Operative technique

To obtain collateral formation in the PCA territory, the classic EDAS procedure described by Matsushima et al[13], was modified. In OA-EDAS procedure, a sig-

moid scalp incision was made along the course of the occipital artery. Occipital craniotomy of 4 by 8cm was made. The dura was incised along the long axis in the middle. The each dural leaves were divided into several dural leaflets, one centimeter apart, and then infolded into the subdural space, leaving the prominent meningeal artery intact. The arachnoid membrane over the cortical sulci was dissected as widely as possible to promote the in-growth of neovasculature. The occipital artery with its attached galea was placed on the exposed surface of the brain, and was sutured to the margin of dura opening. The bone flap was replaced, which was trimmed at both inlet and outlet of the vascularized flap, and carefully secured to avoid compression of the occipital artery. Finally the scalp flap was closed layer by layer.

Results

Table 1 presents the clinical features of our patients. The age at the time of operation ranged from 2 to 9 years (mean 6.3 yrs). Five patients were boys and 11 were girls. PCA symptoms, such as visual TIAs(Blindness or hallucinations) or visual field defects, were observed in 6 patients (37.5%), whereas 10 of the 16 patients (62.5%) had no visual symptoms.

The preoperative MRI scans demonstrated evidence of PCA territory infarction in 44% (7 of 16) of the patients. Fifty-six percent (9 of 16) of the patients exhibited no PCA territory infarction. And preoperative angiography revealed that all of the hemispheres were in angiographic stage 3 or 4, while angiography demonstarated mild PCA stenosis (less than 50% of the diameter) in 25% (4 of 16) and severe stenosis in 75% (12 of 16) of patients. Preoperative SPECT scanning revealed areas of perfusion defects in 31% of the patients and regions of decreased perfusion in 87.5%. Acetazolamide SPECT scanning demonstrated decreased reserves in 94% (15 of 16) of the patients.

The surgical outcomes are summarized in Table 2. The PCA territory symptoms were well controlled in 88% (14 of 16) of the patients, but persisted in two patients with an established visual field defect. The most common MRI finding was of no additional infarction after surgery. Eighty-one percent of the patients who underwent follow-up MRI exhibited favorable MRI changes.

Most patients exhibited good or fair revascularization of the PCA territory (87.5%), however only 8 patients (50%) underwent follow-up angiograms. On the basis of the changes in the SPECT findings for the PCA territory after surgery, most of the patients demonstrated a favorable outcome. Favorable changes were present in 13 of the 16 PCA territories (81%) (Fig. 2).

Post-operative infarctions of variable sizes were the most common complication[11], but in this series post-operative infarction occurred in only one patient (6%). Post-operative epidural hematoma(EDH) or subdural hematoma(SDH) occurred in 3 patients, and post-operative TIAs were observed in 3 patients. All of the patients with both EDH(or SDH) and TIAs recovered normal function without any neurologic deficits or TIAs before discharge.

Table 1. Clinical features of the 16 patients with Moyomoya disease*

Clinical features (n = 16)	No. of Cases	(%)
Age	Mean 6.3 yr	
0~5 yr	5	(31 %)
6~10 yr	11	(69 %)
Sex		
Male	5	(31 %)
Female	11	(69 %)
Clinical manifestation : Visual Symptoms		
No symptom	10	(62.5%)
Visual TIA	4	(25 %)
Visual field defect	2	(12.5%)
MRI findings : PCA territory		
No Infarction	9	(56 %)
Infarction	7	(44 %)
Angiographic Stages**		
III	8	(50 %)
IV	8	(50 %)
Angiographic feature : PCA Stenosis***		
Mild	4	(25 %)
Severe	12	(75 %)
SPECT findings		
Perfusion defect	5 / 16	(31 %)
Decreased perfusion	14 / 16	(87.5%)
Decreased reserve	15 / 16	(93.7%)

* TIA, transient ischemic attack ; MRI, magnetic resonance imaging ; SPECT, single-photon emission computed tomography ; PCA, posterior cerebral stenosis ** angiographic stages were evaluated according to the criteria described by Suzuki and Kodama[19] *** mild stenosis , less than 50% of diameter ; severe stenosis, more than 50% of diameter

Table 2. Surgical outcomes

Parameters	No. of cases (%)	Mean follow-up period
Outcome of PCA symptoms (n = 16)		11.8 months
Favorable	14 (88%)	
Unfavorable	2 (12%)	
Neuroimaging (MRI) change (n = 11)		10.1 months
Favorable	9 (81%)	
Unfavorable	2 (19%)	
Extent of revascularization of PCA territory (n = 8)		5.4 months
Good	3 (37.5%)	
Fair	4 (50%)	
Poor	1 (12.5%)	
SPECT changes of PCA territory (n = 16)		10.9 months
Favorable	13 (81%)	
Unfavorable	3 (19%)	

* MRI, magnetic resonance imaging ; SPECT, single-photon emission computed tomography ; PCA, posterior cerebral stenosis

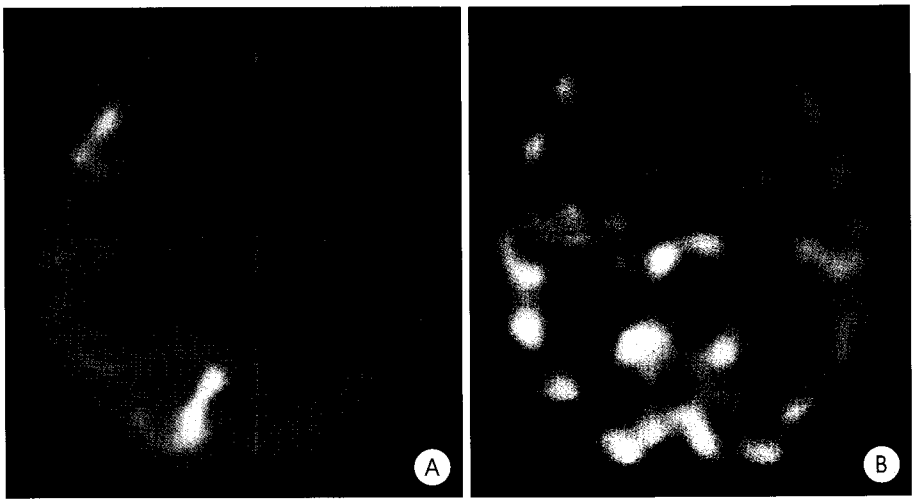

Fig. 2. Acetazolamide single photon emission computed tomographic(SPECT) scans obtained before (A) and after (B) encephaloduroarteriosynangiosis procedure using the occipital artery (OA-EDAS). Disturbed vascular reactivity in response to acetazolamide injection is observed preoperatively in the left occipital lobe (A). Significant improvement of vascular reactivity in reponse to the acetazolamide in the area corresponding to the lesion on preoperative SPECT is observed after OA-EDAS (B).

Discussion

For the revascularization of the PCA territory, several operative approaches, including direct bypass procedures such as OA-PCA[15], indirect bypass procedures such as omental transplantation,[19] burr hole operations[1,9], and other techniques[10,14] have been used as supplementary measures for patients with MMD. The results obtained with these procedures have been reported to be excellent, however it is still unclear which is the safest and most effective. Direct bypass surgery provides immediate improvement of the CBF, but this procedure has technical limitations in the treatment of pediatric MMD, because of the small size of the donor and recipient vessels and the necessity for temporary occlusion of the blood flow in the cortical artery in order to perform direct anastomosis[7]. EDAS using the parietal branch of the STA and STA-MCA anastomosis with encephalomyosynangiosis (EMS) are effective methods of reconstructive vascular surgery for pediatric MMD, but do not always adequately prevent ischemia in the PCA territory, as demonstrated by the CBF[3,11,23]. Consequently, other methods are often necessary to increase the CBF in the PCA territory.

For children with ischemic symptoms, indirect procedures are more often used, because they offer higher rates of successful revascularization and greater technical ease[7]. Omental transplantation has certain advantages, such as the establishment of extensive collateral circulation and the powerful potential of the omental tissue to produce lipid angiogenic factors and endothelial growth factors[4]. However, omental transplantation is relatively invasive in cases of pediatric MMD, because of the need for a laparotomy and a large craniotomy (which potentially disrupts the preexisting collateral vessels). The burr hole operation is a simple and safe procedure, and there are no site limitations because no donor organ is needed[1,9]. However, one disadvantage of this method may be its unpredictable revascularization results, because it is generally accepted that the establishment of collateral circulation requires available tissues capable of furnishing future collateral vessels[17,18].

We selected OA-EDAS as a supplementary measure for the PCA territory, because all of our patients were children and our policy was to produce more predictable collateral formation with minimal risk. By using OA-EDAS, we believed that it would be possible to enhance the collateralization of the ischemic PCA territory.

In most of the patients for whom infarction was observed on MRI, the postoperative MRI scans demonstrated no changes in the previous infarctions, in spite of the surgical treatment. Therefore, early surgical intervention, before the occurrence of definite infarctions, is essential in the treatment of patients with MMD.

It is generally accepted that improvements in the clinical symptoms after surgery become evident before the angiographic appearance of collateral circulation. EDAS has been reported to be an effective approach for the treatment of MMD, in that the TIAs disappeared within 1 year for more than 75% of the patients treated with this modality[22]. In this study, 14 of the 16 patients (88%) showed good clinical improvement. These findings might constitute direct evidence of the resolution of the PCA ischemia. Because angiography is an invasive technique for children, we limited its use to just before the secondary operation and, 1 to 2 years after the last operation. It is difficult to evaluate the visual symptoms with a conventional ophthalmologic exam in children. For more precise evaluation of visual function, the visual evoked potential test is also needed in the case of MMD patients with PCA involvement[22].

SPECT scanning is the most widely used method of evaluating the hemodynamic changes in cases of MMD. The SPECT findings, especially those of hypoperfusion, are generally well correlated with the clinical symptoms[16]. It is known that revascularization develops when the cortex is under hemodynamic stress, which is defined as decreased vascular reserve on preoperative acetazolamide SPECT scans[18]. Good

surgical revascularization provides a significant increase in the resting regional CBF and vascular reserve in the ischemic cortices, and these findings are consistent with clinical improvement[17-19].

We were concerned that the addition of OA-EDAS to STA-EDAS would increase the risk of postoperative ischemia, as compared with simple EDAS, because of the longer operative time and wider surgical field. Therefore, during the OA-EDAS operations, close hemodynamic monitoring was performed, along with aggressive intraoperative management of the blood pressure and adequate partial pressure of CO_2. In this study, postoperative infarction occurred in only one of the sixteen patients. Because impaired cerebral vascular reserve makes patients vulnerable to the hemodynamic stress of surgery, regardless of the surgical modality, patients must be maintained in a normocapneic and normotensive state during the perioperative period[17]. We recommend to keep blood hemoglobin level in normal range, and mild to moderate hypervolemic state for 3 to 5 postoperative days to prevent risk patient from ischemic insult.

Conclusion

OA-EDAS is a safe and an effective revascularization procedure for the PCA territory in MMD patients with compromised PCA territory cerebral vascular reserve, or with visual TIAs of this disorder. Nevertheless, OA-EDAS should be performed to prevent progression of occipital lobe ischemia when the SPECT findings show progressive hemodynamic abnormalities in the PCA territory, even though visual symptoms are absent.

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Commentary

Authors performed occipital artery-EDAS in sixteen patients out of 49 operated cases of moyamoya disease between Aug, 2003 and Apr, 2004. Vascular insufficiency in PCA territory tend to be neglected in treatment of moyamoya disease. Authors analysed the outcome of this procedure successfully on the bases of a clinical course, MRI findings, angiographic changes and SPECT findings. I therefore would like to congratulate the successful outcome of this surgical procedure.

At first, I think that there will be some debate on indication of this procedure. Ten out of sixteen patients had visual sy-

mptoms and the others were selected for surgery from angiographic and SPECT findings. In cases with mild stenosis of PCA with some reduction of hypoperfusion at PCA territory it will be very difficult for neurosurgeons to make decision to have this procedure.

In regards to postoperative evaluations of clinical and MRI findings, it is not easy to confirm as surgery results. In the case that the evaluation of the outcome is favorable, without clinical aggravation or MRI changes could be overestimated simply as a surgical effects, which could similarly occur in non-operated cases.

Authors performed OA-EDAS in conjuction with STA-EDAS and bifrontal EGS, which might result in a long operation time. However I worry about the long operation time in moyamoya disease patients. Fortunately postoperative infarction occured in only one of sixteen patients. But personally, I believe that shorter operation time will be beneficial to reduce perioperative complications. Nevertheless, I would like to give authors high credit for their good trial and excellent outcome.

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