Multiple Concurrent Anastomotic Procedures in the Management of Moyamoya Disease: A case report with review of literature

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ABSTRACT

It has already been proved by many studies that surgical revascularization definitely helps in curing the symptoms of moyamoya vasculopathy. In this regard, we present a case of moyamoya disease which was cured by concurrent multiple anastomotic procedures, namely superficial temporal artery (STA), middle cerebral artery (MCA) anastomosis, encephalomysynangiosis (EMS) and encephalogaleosynangiosis (EGS). A 24-year-old woman presented with symptoms of cerebral ischemia. Thorough investigation with MRA and MRI revealed moyamoya vasculopathy and was confirmed by cerebral angiogram. Multiple concurrent combined anastomotic procedures on both sides relieved the symptoms, which was also confirmed angiographically. A Combination of multiple direct and indirect procedures covers the whole ischemic cortical area and provides effective neovascularization.

Key words: Moyamoya disease, STA-MCA anastomosis, EMS, EGS

"Moyamoya", literally meaning “puff of smoke”, is one of the relatively uncommon central nervous system diseases, mainly found in Japan and other neighborhood countries. Ischemic attack is the commonest presentation and there are various ways of supplying the extra blood flow to meet the demand. Various direct and indirect anastomotic methods are in common practice. Superficial temporal artery (STA) to middle cerebral artery (MCA) anastomosis, STA to anterior cerebral artery (ACA) anastomosis and occipital artery to MCA anastomosis are examples of direct procedures1,2,7,9,10,14,16. Similarly, encephaloduroarteriosynangiosis (EDAS), encephalomyosynangiosis (EMS), encephalomyoarteriosynangiosis (EMAS), encephalogaleosynangiosis (EGS), multiple cranial burr holes, omental transplantation etc are examples of indirect procedures5,8,10,14. Symptomatology, radiological findings and cerebral blood flow are the guidelines for the appropriate anastomosis. Only one or more than one anastomosis can be performed, depending on the extent and severity of cerebral ischemia, to get the optimal effect.

We hereby present a case of moyamoya disease in which three different types of anastomoses, namely STA-MCA anastomosis, EMS and burr hole procedure (EGS) were performed in one stage. This article mainly deals with the surgical techniques used in this particular case and reviews the significance of multiple concurrent anastomotic procedures.

CASE REPORT

A 24-year-old woman presented to our hospital with the main complaints of right partial seizure with numbness of the upper and lower limbs on the right side. She was referred to Hiroshima University Hospital with suspected moyamoya disease on the basis of magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA).

MRI disclosed cerebral infraction on bilateral MCA territory mainly on the left side (Fig. 1a). Single photon emission computerized tomograp-
phy (SPECT) showed bilateral low cerebral blood flow, mainly in the left MCA region (Fig. 1 b). Pre-operative cerebral digital subtraction angiography (DSA) showed typical moyamoya vasculopathy (Fig. 2, 3). There was no evidence of development of any collateral circulation to the ischemic territory from external carotid vessels (Fig. 4). We diagnosed moyamoya disease, and decided to perform bilateral surgical revascularization because of the presenting ischemic events and decreased vascular reserves.

The STA-MCA anastomoses with EMS for the bilateral MCA territories and EGS for bilateral ACA territories were performed in the same stage. After surgery, the previous symptoms disappeared and no other new symptoms appeared. The follow-up angiogram, at 23 months of surgery, showed excellent collateral circulation development in the MCA and ACA territories (Fig. 5).

**SURGICAL PROCEDURE**

i) STA-MCA Anastomosis with EMS

After skin incision, STA and its parietal branch were dissected out from the surrounding tissue. A donor vessel of about 12 cm in length was dissected. The underlying temporalis muscle was elevated from the temporal bone and reflected and a biconvex shaped bone flap was removed with craniotomy. The dura was incised and brain surface exposed. A recipient vessel of appropriate size, M4 segment of MCA, was selected with the help of vascular Doppler. After preparing the donor and recipient vessels, end to side STA-MCA anastomosis was performed (i.e. the end of STA was anastomosed with the sidewall of MCA). Temporary clips were released and good blood flow across the anastomosis was confirmed with the help of vas-
Fig. 3. Pre-operative left vertebral angiogram: a) antero-posterior view, b) lateral view of early phase and c) lateral view of late phase, showing extensive development of moyamoya vessels and posterior pericallosal artery. A large part of cerebral hemisphere is supplied from the posterior circulation through leptomeningeal anastomosis.

Fig. 4. Pre-operative left external carotid artery angiogram (ECA), lateral view: a) early phase and b) late phase, showing no collateral circulation from the branches of ECA.
cular Doppler.

EMS was also performed at the same stage. The ischemic cortical surface on the fronto-parietal region was exposed by extending the dural opening. The temporalis muscle which was reflected before craniotomy was placed over the ischemic cortical surface and the margin of muscle was sutured with the dural margin.

**ii) EGS**

A burr hole was made on the frontal region, a few centimeters lateral to midline and in front of the coronal suture. Dura was exposed and cut in cruciate fashion and the brain surface was exposed. Galeal layer of the reflected skin flap was dissected off from the underlying tissues. The dissected galeal layer was gently inserted over the brain surface through the burr hole and the scalp was closed.

**DISCUSSION**

Surgical management for moyamoya disease has been in the process of development for last more than 50 years\(^\text{10}\). It has already been proved that surgical revascularization definitely helps in moyamoya vasculopathy\(^1,2,6,8,11\). In older days, only one anastomotic procedure used to be performed and in some cases with a wide ischemic region, another type of procedure would be added. However, now the trend of doing multiple combined anastomotic procedures has been developed.

Many studies have evaluated the technical aspects and effectiveness of different approaches to treat cerebral ischemia related to moyamoya vasculopathy. It has been found that the technique of multiple combined procedures is better than a single procedure, and that a direct anastomotic procedure is more effective than an indirect procedure\(^5,6,12,13\). However, the technical difficulties of the direct method, especially in children, and the severity of possible complications, e.g. stroke and intracerebral hemorrhage, sometimes limit this technique when an alternative exists. Studies have shown that indirect methods also have an angiographically, electorencephalographically and symptomatologically significant neovascularizing effect, besides being technically simple\(^3\). At the same time, the drawbacks of indirect methods, mainly the inability to develop sufficient collaterals, also need to be taken into account. EMAS or EMS also tends to cause epidural hematoma and muscle calcifications in the long run\(^16\). In this context, a combination of direct and indirect anastomotic procedures has been found to have a better effect than any single procedure. Multiple combined procedures are also necessary because of the fact that the area covered by each single procedure is not wide enough.

Though other different combinations of anastomotic procedures have been mentioned in literature, the combination of STA-MCA anastomosis, EMS and EGS does not seem to have become a common practice yet. We found that this combination is quite effective and is a simple method. In this particular case, the MCA and ACA territory of left side was ischemic, and to cover the whole ischemic area, multiple procedures were performed. STA-MCA anastomosis and EMS were performed to cover the MCA territory and EGS was performed for ACA territory. Instead, STA-ACA anastomosis, or frontal EMAS using the frontal branch of STA could be an alternative to EGS for ACA territory if the ischemic zone is too wide and severe\(^2\).

STA to MCA revascularization is a procedure to treat selected patients with intracranial ischemia due to morbid stenosis of terminal ICA, which is not accessible by carotid endarterectomy. An extra blood flow to the middle cerebral artery opens the
new collateral channels and improves perfusion by providing extra blood flow to these ischemic areas. The combination of STA-MCA anastomosis and EMS is already an established approach for treating moyamoya disease\(^\text{4,12}\).

EGS is also one of the simple yet effective, indirect anastomotic procedures provided the ischemic zone is not too wide. In this procedure, the extra blood flow is provided to the cerebral cortex from the galeal layer of the scalp. Therefore, this procedure is called encephalogalectosynangiosis (EGS). Though EGS alone is less commonly performed as a main anastomotic procedure, it has been found to be an important supplement to other anastomotic procedures. Therefore, the combination of above mentioned procedures was found to be not only technically simple, taking hardly about 5-6 hours for the complete surgery, but also effective as evidenced by post-operative evaluations. Though the direct method is more effective, the indirect method is safer, and thus combination of both gives a more effective and safer outcome. Studies have demonstrated that a combination of direct and indirect anastomosis resulted in significant neovascularization in moyamoya disease\(^\text{4,12,13}\).

In the present case, as, shown in Figure 5, the post-operative angiography after 23 months of surgery showed a significant neovascularization and thus no subsequent recurrence of symptoms. Though there are some controversies regarding which procedure, direct or indirect anastomosis, is better, a combination of both in a single stage is the best approach according to current literature and our experience.

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