Interdisciplinary Neurosurgery 14 (2018) 102–110

Contents lists available at ScienceDirect

Interdisciplinary Neurosurgery

journal homepage: www.elsevier.com/locate/inat

Unilateral moyamoya disease mimicking intracranial hemorrhage in a pediatric patient: Surgical treatment with encephalo-duro-myo-synangiosis in a progressive disease

Ahmad Faried*, Rhonaz P. Agung, Yogi Rosbianto, Mirna Sobana, Muhammad Z. Arifin

Department of Neurosurgery, Faculty of Medicine, Universitas Padjadjaran–Dr. Hasan Sadikin Hospital, Bandung, West Java, Indonesia

ARTICLE INFO

Keywords:
Unilateral moyamoya disease
Encephalo-duro-myo-synangiosis
Ischemia

ABSTRACT

Background: Moyamoya disease (MMD) is characterized by progressive bilateral internal carotid artery stenosis and its distal branches. When angiographic findings are unilateral, the diagnosis is considered to be probable MMD. Surgery is recommended treatment of patients with deterioration caused by progressive cerebral ischemic events. Different techniques have been described, and the encephalo-duro-myo-synangiosis (EDMS) procedure is highly recommended for patients with rapid deterioration. A case of unilateral MMD mimicking intracranial hemorrhage in a pediatric patient is reported.

Case description: A 14-year-old girl was referred to our center due to a decreased level of consciousness. Physical examination revealed right-sided hemiparesis; however, a brain CT scan showed multiple hyperdense masses mimicking a hemorrhagic lesion. A brain MRI showed a large hypointensity in the temporoparietal, suggesting a vascular lesion. In all children that exhibit encephalitis, vascular events such as MMD should be considered. An emergency surgical EDMS procedure was performed, and the patient regained consciousness and exhibited no TIAs during the follow-up period. A postoperative brain MRI showed an improvement in brain vascularity.

Conclusion: After EDMS, a considerable degree of neurological recovery was observed in our patient with rare unilateral MMD mimicking intracranial hemorrhage. We found that the progression of clinical improvement after indirect revascularization in our case was due to EDMS. Surgical treatment with EDMS is reasonable for MMD because it allows for flexible revascularization that adequately addresses the requirement for new blood supply in the ischemic lesion. The EDMS procedure is followed by an observation period to preclude the possibility of the patient developing definite MMD.

1. Introduction

Moyamoya disease (MMD) is an unremitting cerebrovascular occlusive disorder of unknown etiology that is becoming more widely recognized as a cause of stroke in pediatric patients. It is characterized by progressive occlusion of both the internal carotid artery (ICA) or it's terminal branches, along with the formation of an extensive vascular network known as moyamoya vessels. When the angiographic imaging results are unilateral, the diagnosis is classified as probable MMD. In Japan, definite (bilateral) MMD is the most common pediatric cerebrovascular disease and affects females almost twice as often as males [1]. Unilateral MMD is a relatively rare disease, and the prevalence was 10.6% of 2635 patients with MMD in Japan [2].

Surgical treatment is recommended for patients with recurrent or progressive cerebral ischemic events and associated reduced cerebral perfusion reserve [1, 3]. Many different surgical techniques have been described, and all have a main goal of preventing further ischemic injury by increasing collateral blood flow to hypoperfused areas of the cortex by using the external carotid circulation as a donor supply. This article reports our use of an encephalo-duro-myo-synangiosis (EDMS) procedure, which is a method of indirect revascularization, in a patient with unilateral MMD. The EDMS procedure is a safe, effective, easy and durable method of cerebral revascularization for MMD and should be considered a primary treatment for moyamoya, particularly if the

Abbreviations: MMD, moyamoya disease; ICA, internal carotid artery; EDMS, encephaloduromyosynangiosis; RSHS, Dr. Hasan Sadikin Hospital; CT, computed tomography; MRI, magnetic resonance imaging; MRA, magnetic resonance angiography; MCA, middle cerebral artery; CSF, cerebrospinal fluid; TIAs, transient ischemic attacks; ECA, external carotid artery; STA, superficial temporal artery; EDAS, encephaloduroarteriosynangiosis; EMS, encephalomyosynangiosis

* Corresponding author at: Department of Neurosurgery, Faculty of Medicine, Universitas Padjadjaran–Dr. Hasan Sadikin Hospital, Jl. Pasteur No. 38, Bandung 40161, West Java, Indonesia.

E-mail address: ahmad.faried@unpad.ac.id (A. Faried).

https://doi.org/10.1016/j.inat.2018.07.008
Received 23 May 2018; Received in revised form 2 July 2018; Accepted 22 July 2018

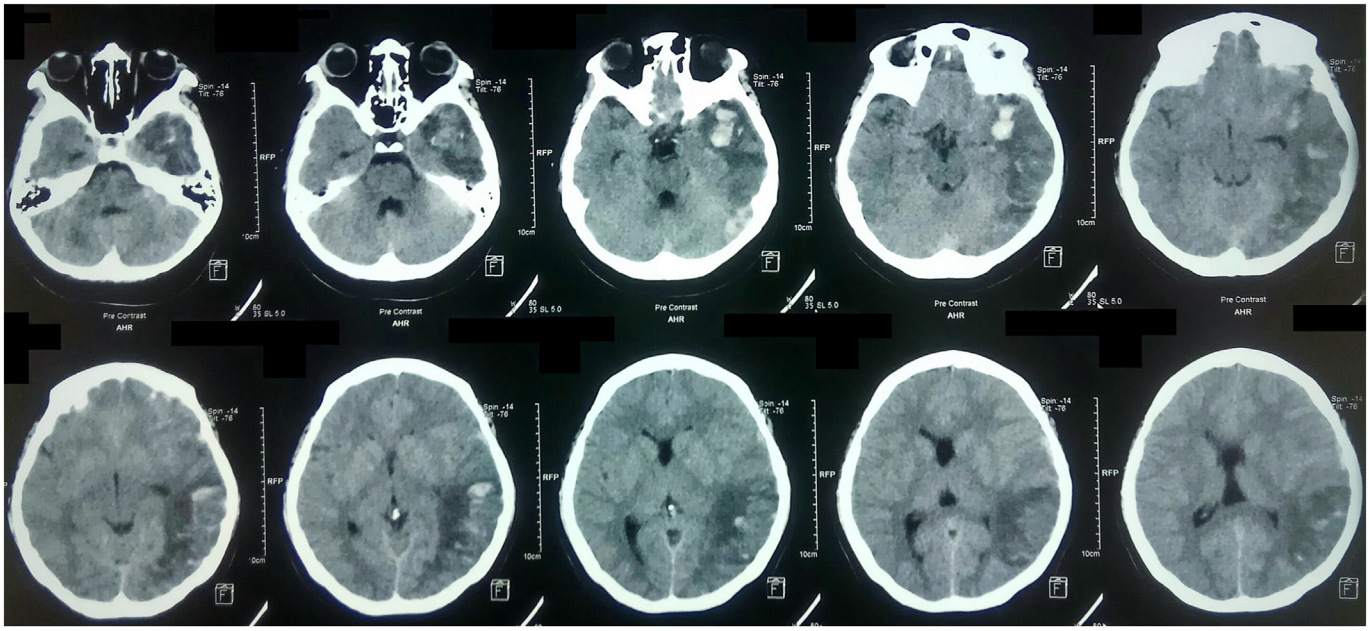

Fig. 1. The patient was misled by hemorrhage lesion because of CT Scan shown multiple hyperdense mass at temporoparietal.

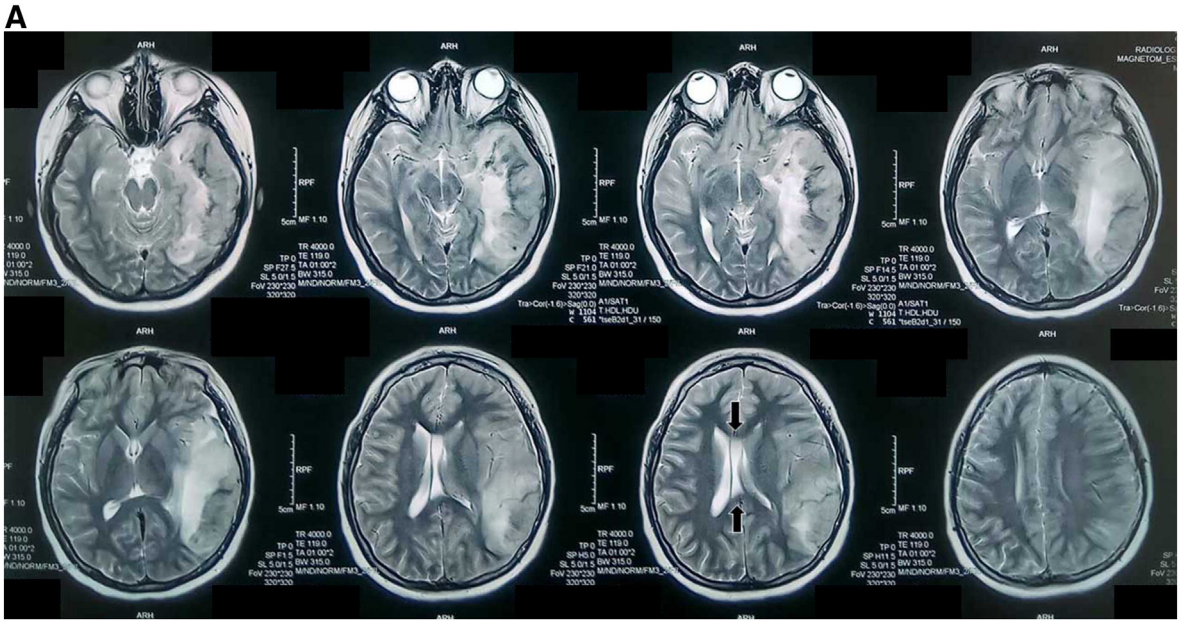

Fig. 2. Considering the ischemic symptoms, we than performed other examinations using magnetic resonance-imaging (MRI), that showed (A) the presence of a wide hypointense lesion at left temporal and parietal lobes intermingled with brain parenchyma without a clearly identifiable margin in T2-Weighted; as coexistent of cavum septum pellucidum and cavum vergae shown with black arrow, (B) MRI Diffusion-Weighted imaging revealed diffusion restricted in the left MCA territory, and (C) MRI Apparent Diffusion Coefficient showing dark signal corresponding to area of large infarct.