

Case report

Association of mesial temporal sclerosis and moyamoya syndrome

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1. Introduction

The moyamoya phenomenon is a progressive intracranial arteriopathy with concomitant formation of fragile collateral vessels, leading to ischemic and hemorrhagic events [1]. Moyamoya disease specifically refers to idiopathic, bilateral disease. Unilateral idiopathic disease or arteriopathy in association with known medical conditions is termed moyamoya syndrome [1]. Moyamoya syndrome has been associated with a history of cranial radiation, neurofibromatosis, Down syndrome, sickle cell disease, congenital cardiac anomalies, renal artery stenosis, and hyperthyroidism [1]. To our knowledge, the association of moyamoya syndrome and mesial temporal sclerosis has not been well-reported.

2. Case presentation

This 29-year-old right-handed Caucasian female presented to our institution with an 11-year history of complex partial seizures with rare secondary generalization. The seizures were characterized by an initial subjective sensation of feeling hot followed by an arrest in speech. Some were also characterized by lip automatisms preceding the speech arrest. Her seizures had generalized

three times, resulting in brief tonic–clonic episodes. These seizures were localized to the left temporal region via EEG demonstrating occasional sharps from this region. In retrospect, she did recall one particular atypical event 8 years prior characterized by a transient peripheral vision disturbance followed by possible hemiparesis and syncope. MRI demonstrated increased mesial temporal FLAIR signal (Fig. 1A) and PET imaging demonstrated decreased metabolism in this region as well (Fig. 1B), consistent with a diagnosis of mesial temporal sclerosis. Careful perusal of the original MRI also demonstrated relative stenosis of the ipsilateral supraclinoid ICA, A1 and M1 (Fig. 1C). Subsequent formal digital subtraction angiography performed for Wada (intracarotid sodium amobarbital) testing confirmed moyamoya vasculopathy in the left hemisphere (Fig. 1D and E).

Given the suspicion of a history of possible TIAs and risk for further ischemic events, the patient was taken to the operating theatre for STA to MCA bypass. After identifying the course of the STA via Doppler, a small curvilinear incision was performed over its course under the operating microscope. After isolation of the artery, the temporalis muscle was opened and a small temporal craniotomy was turned. After opening the dura, the arachnoid over the recipient M4 vessel was dissected. The STA was then sutured to the recipient MCA branch with an end-to-side anastomosis. The procedure was uncomplicated and postoperative CTA confirmed patency of the graft (Fig. 1F). At 1.5 year follow-up, she has not had any events suspicious for ischemia and her seizure frequency has improved with medical management.

3. Discussion

Moyamoya collateralization may develop as a result of ischemic disease such as atherosclerosis as well as congenital disease such

Abbreviations: ICA, internal carotid artery; A1, anterior cerebral artery, first segment; M1, middle cerebral artery, first segment; STA, superficial temporal artery; MCA, middle cerebral artery; CTA, computed tomography angiography; β FGF, beta fibroblast growth factor.

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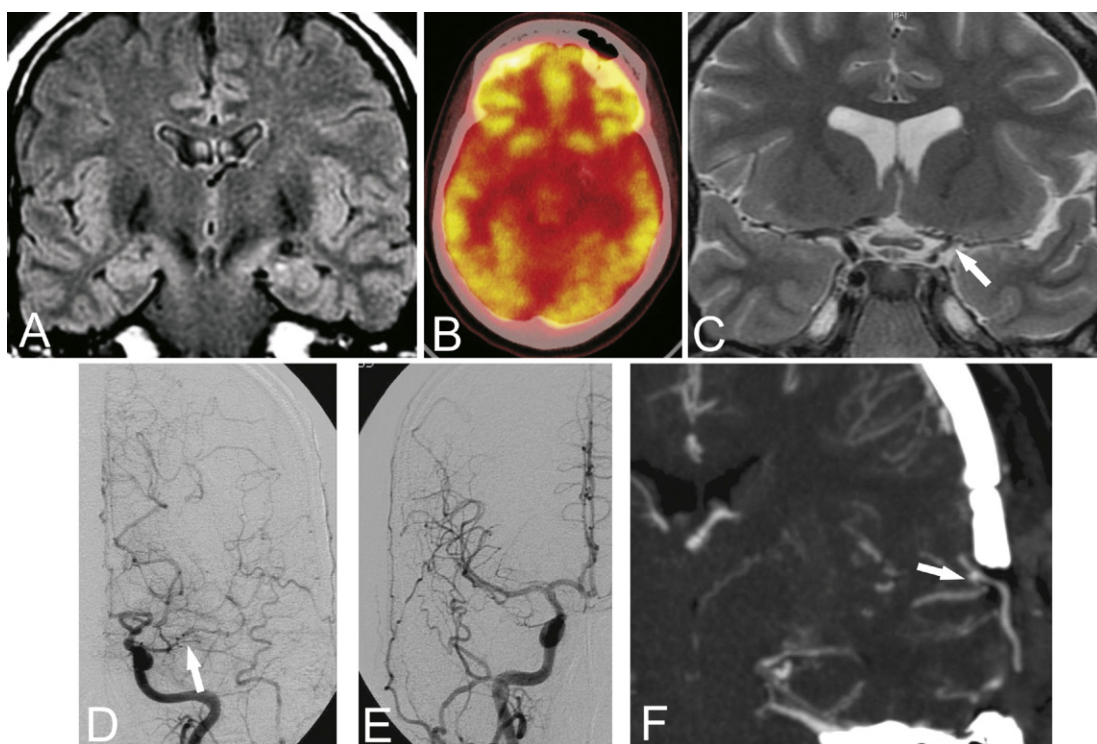


Fig. 1. This 29 year old Caucasian female presented with an 11 year history of complex partial epilepsy. Coronal T2-weighted FLAIR MRI demonstrated increased signal in the left mesial temporal lobe (A), and axial PET scan demonstrated decreased metabolism in this region (B). In retrospect, MRI also demonstrated stenosis of her ipsilateral supraclinoid ICA, A1 and M1 branches (arrow, C). Digital subtraction angiography performed for WADA testing confirmed this stenosis and demonstrated moyamoya collateralization (arrow, D). Vasculopathy was not appreciated on digital subtraction angiography of the contralateral side (E). The patient underwent successful STA–MCA bypass, confirmed by postoperative CTA (arrow, F).

as sickle cell disease [1]. It may then serve as a source of subsequent ischemic and/or hemorrhagic events [1]. Interestingly, a component of the pathophysiologic mechanism of mesial temporal sclerosis is also ischemia [2,3]. This pathophysiologically links these two diseases and may explain their concomitant development in this patient. It is important to reiterate that the vessel stenosis and moyamoya collateralization was indeed ipsilateral to the mesial temporal sclerosis in this patient, suggesting a relationship between them rather than coincidence. This relationship may be the result of a unifying ischemic event that resulted in the development of both disease processes, or alternatively, mesial temporal sclerosis may have occurred as a result of ischemia from unilateral moyamoya syndrome. The latter may explain the mechanism of seizure presentation for some patients with moyamoya syndrome [1] and also reinforces the etiologic role of ischemia in mesial temporal sclerosis [2,3]. A deeper pathogenetic link between these two phenomena at the molecular level may lie in increased expression of β FGF in patients with temporal lobe epilepsy and seizures [4]. Interestingly, β FGF is also known to stimulate endothelial cell growth and promote angiogenesis, and is well known to be elevated in patients with moyamoya [5].

Regardless of the exact mechanism of association, this report encourages careful examination of vessels ipsilateral to mesial temporal sclerosis, either on T2-weighted imaging or, ideally, with formal digital subtraction angiography for WADA testing. In this case, as a result of detection of moyamoya syndrome, we were able to successfully augment flow to the patient's symptomatic hemisphere. She has since had no ischemic events in 1.5 years of follow-up and her seizure frequency has declined.

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