Case Report / Olgu Sunusu

Choreiform movements due to pediatric Moyamoya disease - A case report with a dramatic response to therapy
Pediatrik Moyamoya hastalığına bağlı koreiform hareketler- tedaviye dramatik cevap veren bir vaka sunumu

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Başvuru Tarihi/Received :
16-09-2013
Düzeltme Tarihi/Revised:
01-12-2013
Kabul Tarihi/Accepted:
22-12-2013

ÖZET

Moyamoya hastalığı internal karotid arterin terminal bölümü ile ana dallarının ilerleyici stenozu ile karekterize sık görülmeyen bir serebrovasküler bir hastalıktır. Geçici iskemik atak, iskemik inme, intrakraniyal kanamalar, nöbetler, baş ağrısı, koreiform hareketler ve kognitif fonksiyon bozukluğu gibi çeşitli semptomları vardır. Burada geçici iskemik atak, sol hemiparezi ve tek taraflı koreiform hareketleri olan 14 yaşına moyamoya hastalığı tanısı konulan bir hasta sunuldu. Koreiform haraketler, ensefaloduroarteriyosinanjiozis operasyonu sonrası iki taraflı ve aşrırı şiddetlendi. Bu şiddetli koreiform hareketler dopamin reseptör blokör tedavisine dramatik cevap verdi.

Anahtar kelimeler: Arteriyovenöz malformasyon, kore, moya moya hastalığı

ABSTRACT

Moyamoya disease is an uncommon cerebrovascular disease that is characterised by progressive stenosis of the terminal portion of the internal carotid artery and its main branches. There are a multitude of symptoms associated with Moyamoya disease, including transient ischaemic attacks, ischemic strokes, intracranial hemorrhages, seizures, headaches, choreiform movements, and cognitive deficits. We report here a 14-year-old male child with Moyamoya disease who presented with overlooked transient ischaemic attacks, left-sided hemiparesis, and unilaterally choreiform movements that occurred bilaterally after surgery. The latter gave a dramatic response to dopamine receptor blocker therapy.

Key words: Arteriovenous malformation, Chorea, Moyamoya disease

Introduction

Moyamoya disease (MMD) is a disease of unknown origin characterized by progressive stenosis and, ultimately, occlusion of the distal intracranial internal carotid arteries (ICA) and the proximal branches of the anterior and middle cerebral arteries. The most common presenting feature in pediatric MMD is cerebral ischemia; in contrast, choreiform movements which is less common in children (1, 2).

We describe a case with MMD who admitted unilaterally choreiform movements and became bilaterally after encephaloduroarteriosynangiosis surgery that responsed to dopamine receptor blocker therapy quickly.

Case Report

A 14-years-old boy presented with left-sided hemiparesis and choreiform movements for 6 months. In his history, there were recurrent episodes of headache and three episodes of left-sided hemiparesis lasting 2-3 days following headache lasting for a half for two years which was a transient ischemic attack.

Clinical examination showed left-sided hemiparesis, positive babinski sign and increased deep tendon reflexes, and choreiform movements on the upper left side extremity. He had the clinical features of neither Down syndrome nor neurofibromatosis. Complete blood count, biochemical tests, electrolytes were unremarkable. Hematological consultation was normal for sick cell anemia.

Computed tomogram brain was normal. Magnetic Resonance Imaging showed in T1 weight image, heterogeneous contrast increments observed in vascular collaterals at the level of bilateral basal ganglia and talamus formed by talamoperforan and lenticulostriate arteries

narrowing of left middle cerebral artery and lot of collaterals. Further, digital subtraction angiography was done and it showed stenosis of left middle cerebral artery and smaller caliber of whole of the left internal carotid artery and vasculer collatarels, stenotic and occlusive arteries were shown and thin vascularization like retikulated was seen in areas supplied by arteries in early arterial phase;(puff of smoke image) (Figure).

MMD was diagnosed and the child was started aspirin (80 mg/day). Treating the child was used the encephaloduroarteriosynangiosis (EDAS) as a neurosurgical procedure. One week after the operation, choreiform movements increased and became bilaterally and he was not able to eat his meal by himself. The patient was started dopamine receptor blocker (haloperidol) of 1 mg/day, increased the dose up to 3mg/ day, and choreiform movements stopped at the dose. The child is being followed up.

Discussion

MMD was first described in the Japanese literature in 1957 by Takeuchi and Shimizu. In 1969, however, Suzuki and Takaku coined the term “moyamoya” signifying “something hazy, like a puff of cigarette smoke” to describe the angiographic appearance that would both describe and define the illness (3). MMD is seen throughout the world although initial descriptions and studies initially centered on Japanese populations. It affects both children and adults of various ethnic backgrounds (3). MMD accounts for approximately 6% of all causes of pediatric ischemic stroke, although this figure is higher in Japan (4).

There are a multitude of symptoms associated with Moyamoya disease, including transient ischemic attacks, ischemic strokes, intracranial hemorrhages, seizures, headaches, choreiform movements, and cognitive deficits (3). Children with MMD usually present with transient ischemic attacks or cerebrovascular stroke and adult patients present with hemorrhage (5). In the history of our patient, there were recurrent episodes of headaches and three episodes of transient ischemic attack. His parents, however, did not take him to a medical center until choreiform movements emerge. Choreiform movements are seen in 3–6% of patients with MMD, and are attributed to any

dysfunction of the basal ganglia-thalamocortical circuits, including infarctions and mechanical compression by traversing dilated collateral vessels (6). Administration of dopamin receptor antagonists were resolved the choreic movements (7, 8). Our case had unilaterally choreiform movements and furthermore they increased and affected both upper extremities interestingly during the postoperative period. They dramatically responsed to dopamin receptor blockage.

MMD should be considered and worked up in any child presenting with ischemic symptoms, especially in the setting of hyperventilation, crying, and/or physical exertion. A suspected diagnosis of MMD is confirmed with radiological studies. Gold-standard for the diagnosis of MMD is cerebral angiography. Our case typically showed both clinical features and radiological studies for MMD.

Currently, there is no definitive medical treatment for MMD. Antiplatelet agents (*e.g.*, aspirin) and calcium channel blockers may play adjuvant role in the treatment of MMD. Aspirin are usually given to prevent clots. Because a proportion of the ischemic symptoms associated with MMD have been attributed to microthrombus formation due to emboli arising from sites of arterial stenosis (3). However, surgery is usually regarded as the major treatment option for MMD patients suffering from progressive neurological symptoms (3). EDAS, (Pial Synangiosis) was first described by Matsushima, et al (9). Superficial temporal artery is sewn to the inside edge of the dura such that it remains in contact with the exposed cortex. Over time, angiogenesis results in the formation of small arteries to the brain (9). Nerological deficit and morbidity is rarely occurred after EDAS (10). Episodes of tremors with severe headache and simple partial seizure after surgery which resolved spontaneously has been reported as a postoperative complication after EDAS (11). Our patient had had the EDAS neurosurgical procedure and he has been taking an aspirin for one and a half years. He never expericienced any transient ischemic attacks or ischemic strokes and episodes of headache.

The natural history of this disorder is not well known. If left untreated, MMD progresses, and frequently results in permanent neurological and cognitive deficits. Rapid diagnosis of the pediatric patient with MMD is essential, as neurological status at the time of treatment is more predictive of long-term outcome then age. Associated clinical conditions and syndromes should be assessed for, as they are a risk for both the development of MMD and its progression (12). Although there is no curative medical treatment for MMD, numerous studies have shown long-term improvement in children undergoing surgical revascularization procedures (MMD) (3, 13) .

Finally, despite being relatively uncommon, MMD is becoming more widely recognized worldwide as a cause of pediatric cerebrovascular events, and should be considered in any child who presents with symptoms of cerebral ischemia.

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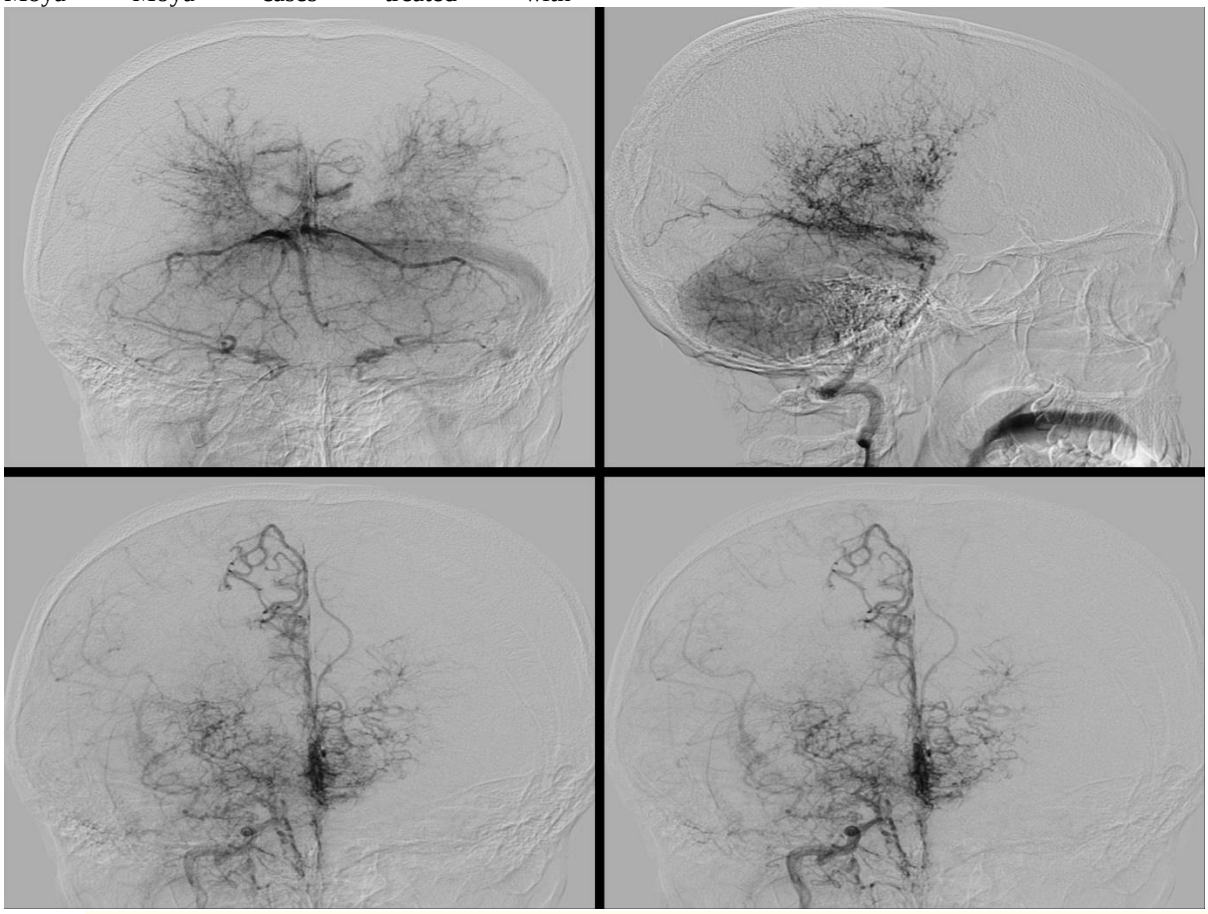

Figure. Digital subtraction angiography showed stenosis of left middle cerebral artery and smaller caliber of whole of the left internal carotid artery and vasculer collatarels, stenotic and occlusive arteries were shown and thin vascularization like reticulated