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Review Article

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Moyamoya Disease: The Intersection of Medical Illness and Mental Distress in Children and Adolescents

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Abstract

Moyamoya disease (MMD) is a progressive vascular disorder in where the carotid artery becomes narrowed, reducing blood flow to the brain. This most commonly presents as stroke in children and young adults, and commonly occurs between 5 and 10 years of age in children and between 30 and 50 years of age.

In MMD survivors, the mental disorders are of highest prevalence which include depression, anxiety, and post-traumatic stress disorder (PTSD). According to studies, anxiety and PTSD are associated with neurological disability, while depression and anxiety are associated with greater cognitive deficiency. The purpose of this case is to increase awareness of an unusual presentation of MMD as psychiatry disorder. It is important for early diagnosis and timely intervention.

Keywords: Moyamoya Disease, Ptsd, Vasculopathy, Depression

Introduction

Moyamoya disease (MMD) occlusive disorder of the cerebral vasculature [1]. There is a progressive stenosis of terminal parts of bilateral internal carotid arteries and the main trunks of anterior and middle cerebral arteries, resulting in the formation of collateral vessels at the base of the brain. These small collateral vessels create "puff of smoke" appearance on angiogram. There are various clinical manifestations for MMD. There is bimodal age presentation with first peak occurring in the first decade of life, associated with cerebral infarction as progressive carotid occlusion develops [1].

The clinical signs of MMD may present itself as cerebral hemorrhage and cerebral ischemia [1]. Both the symptoms present differently in adult and child population. The adult patient most often present in the fourth decade with intracranial hemorrhage arising from rupture of the delicate network of collateral vessels which is mostly intraparenchymal and may be intraventricular or occasionally subarachnoid bleed. In children, the most common presentation is that of recurrent episodes of cerebral ischemia manifesting clinically as focal deficits, paresthesia, and seizures [1,2]. Mental decline maybe the first and sometime the only symptom in children [1,2].

In MMD, MRI and MRA showed bilateral loss of normal flow signal with cavernous and supra-clenoid segments indicating internal carotid artery occlusion [3]. Electroencephalogram (EEG) showed paroxysmal spikes and wave's discharges from left frontal-temporal leads with generalization [3,4].

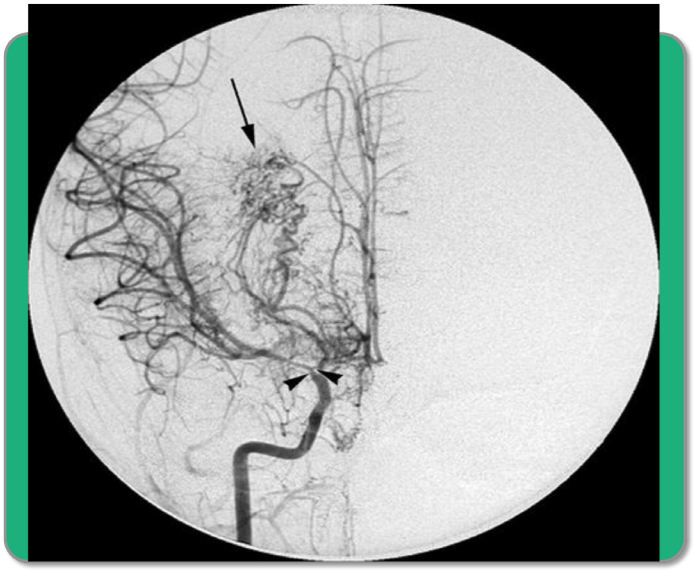

Figure 1: Puff of smoke sign

Clinical features include impairments in mood (e.g. depression, anxiety and PTSD) are associated with cognition (e.g. impaired memory, executive functioning, attention, epilepsy). Although the MMD is the most common in Japan, many subsequent cases have been reported elsewhere, including North America, Europe, and India [4]. Information about the clinical features and long-term outcome of MMD in is lacking. MMD is an easily overlooked condition because of its low incidence and may not be considered as a possible diagnosis for the clinical presentation. Because of its progressive nature, it is imperative to diagnose MMD early and offer surgical treatment [4-6].

Case presentation

A 16-year-old Dominican boy presented to the psychiatric emergency department due to mood dysregulation and aggression. He had medical history of Moyamoya disease, Sickle cell disease, Frontal lobe epilepsy, and cerebrovascular accident in 2019.

Apparently, a year ago he moved from Dominican Republic to USA and attends bilingual regular education classes. He was apparently alright till 2019, then he experienced a cerebrovascular accident/stroke learned of his diagnosis and underwent immediate neurosurgical intervention. His home medications include Aspirin, Valproate, and Hydroxyurea.

He reported recent aggression, and states he started "beating his brother and would not stop" in the context of a phone disagreement. He stated he often had terrible headaches and his aggression worsens. His mother stated that he has been acting aggressive and smiling in the next moment as if nothing happened. He became tearful when describing psychosocial stressors related to his medical illness, his exuberant will to live, fear of death, and family financial stress in the midst of COVID19 pandemic. These factors worsened his anxiety and depression in the last few months. He wanted to die and was tired of living.

According to collaterals, he displayed increased anger outbursts since COVID-19 pandemic. This is related to increasing isolation at home, mild anhedonia, poor appetite, and the exacerbation of anxiety of contracting the virus for fear of death. He does not express his feelings at home, and his family did not know the level of distress he was experiencing. On evaluation, he was cooperative and refused any suicidal, homicidal ideations and no auditory, visual hallucinations.

The patient was diagnosed with Mood disorder due to medical condition. His treatment plan included continued out patient monitoring with a psychiatrist and continuation of medication.

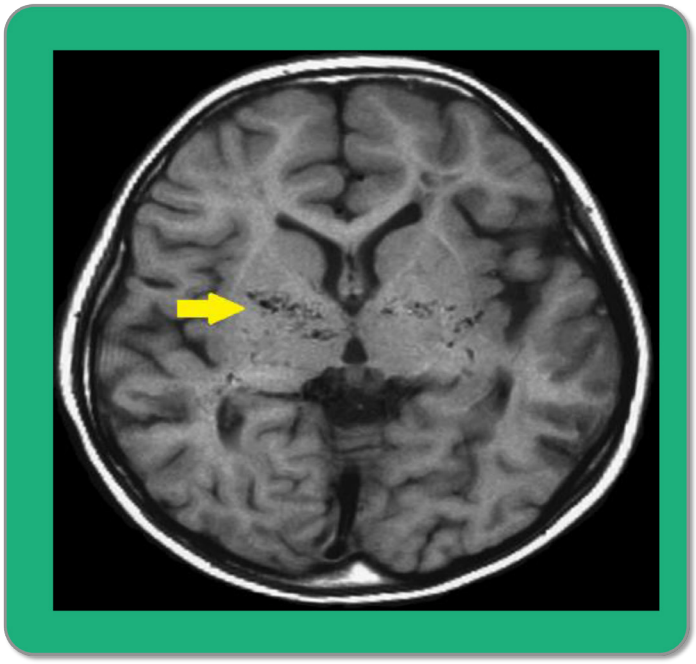

Figure 2: Devoid of blood supply (at arrow)

Disscussion

The exact pathogenesis of MMD is not clear [6]. But there are lot of risk factors that are associated with it like pediatric age group, East Asian genetics, family history of MMD and female gender[1,6]. There are some medical conditions that are also associated with MMD like Downs syndrome, systemic lupus erythematosus, neurofibromatosis, periarteritis nodosa, thyrotoxicosis and previous radiation therapy [7].

Only psychiatric presentations of MMD are rare in literature [8]. The clinical symptoms of MMD include ischemic stroke, transient ischemic attacks, hemorragic stroke, epilepsy, headache, cognitive dysfunction and every symptom varied depending on the age [9]. In adults, anxiety and depression along with focal neurogical deficits is more common. In pediatric patients, hyperactivity, impaired concentration, anxiety, mood lability and poor academic performance have also been described. The clinical signs and symptoms vary, although psychosis appears to be more common in children and adolescents [1,9,10].

This case presented with signs of MMD with psychiatric manifestations which can generally be observed in children but often ignored. It had the risk factors which are often seen with MMD as mentioned above.

Among children with MMD, 20%suffer from headache [1]. It was reported by Seol et al that 44 of 204 children with MMD complained of headache [11]. To diagnose MMD a specific set of criteria needs to be met. Even though which includes occlusive changes in the internal carotid artery (ICA) bilaterally or unilaterally as well as abnormal collateral blood vessels based on radio-

logical imaging. MRI, MRA and CT angiography are the best and practical diagnostic tests but Catheter angiography is the definitive diagnostic tests. But they are less sensitive and have difficulty in detecting basal collaterals [12].

Medical management of psychiatric symptoms of MMD includes mood stabilizers, antidepressants, antipsychotics, benzodiazepines [13-18]. Moyamoya disease associated depression, anxiety responds well to antidepressants and given their relatively safe side effect profile early treatment is recommended. For short term management of anxiety, agitation benzodiazepines appear as a safe option but their efficacy as a monotherapy has been reported to be poor and should be avoided in children [13,16,18]. The incidence of MMD exhibits wide ranging regional differences, with a high incidence in East Asia and low incidence in other regions. Worldwide, MMD age of onset is bimodal in distribution, major peak in first decade of life and moderate peak in the late 20 to 30 years [1,19,20].

Disease progression can occur slowly with occasional stroke or rapidly with advanced neurologic decline. The neurologic status at the time of surgical management is the most important factor that predicts the outcome of the patient both children and adults. Even among asymptomatic patients, the rate and severity of disease progression is high without surgical intervention [21,22]. Conservative therapy includes anti-platelets, statins, aspirin which are not that efficient [21]. A prospective study in Japan estimated a natural progression rate of approximately 20% over 6 years. In symptomatic patients, surgical revascularization is the treatment to prevent cerebral infarction and restore perfusion and reserve capacity [23]. If a major neurological event occurs like stroke or bleeding, irrespective of treatment patient may experience a permanent loss of function with poor prognosis. Therefore, early intervention and prompt diagnosis of MMD is important for achieving better disease outcomes.

Conclusion

The association between MMD and neuropsychiatric behavioral changes including anxiety and PTSD symptoms is acknowledged.

Regarding Moyamoya disease and neuropsychiatric illness, as more robust assessments emerge, it is becoming clearer that the neuropsychopathology of survival from MMD is complex, and multifaceted.

The most obvious alternative to prevent MMD and neuropsychological illness is, in early recognition and surgical correction of narrowing vessels in brain, to prevent repetitive neuropsychiatric complications from occurring later in the life. It is also important to identify and treat psychiatric symptoms to improve quality of life in patients with MMD.

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