CASE REPORT

Moyamoya Disease—"A Puff of Smoke": A Rare Pediatric Case Report

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Abstract

Introduction: Moyamoya disease (MMD) is a rare neurovascular disorder that is characterized by chronic progressive stenosis at the apices and bilateral occlusion of the intracranial internal carotid arteries. The occlusion is gradual in manner; multiple anastomoses are formed between the internal and external carotid arteries. The incidence of MMD has increased in Asian countries with a decline in Europe and United States.

Case description: The case report describes the specific clinical features and dental treatment in a 12-year-old male presenting with MMD with right-side hemiplegia. The patient presented with broken tooth in the upper front tooth region due to trauma. The dental treatment plan focused on providing restorative as well as preventive care to the patient.

Clinical significance: The patients with MMD have an increased risk of stroke development, which may be aggravated by pain or anxiety. It is necessary to avoid cerebral ischemic attacks caused by fluctuating BP and respiration. It becomes imperative to maintain normocapnia, normotension, and pain control during the procedure in order to prevent postoperative ischemic complications. Dental professionals must attain a complete knowledge of this disease and perform the dental treatment in pain and stress-free manner.

Keywords: Dental treatment, Ischemic stroke, Moyamoya disease, Trauma.

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Introduction

Moyamoya disease (MMD) is a cerebrovascular disorder due to chronic stenosis at the apex of the intracranial internal carotid arteries, which includes proximal anterior cerebral arteries and middle cerebral arteries and bilateral occlusion of the internal carotid arteries.[1]

The word "moyamoya" is from Japanese origin, meaning "something hazy" like a "puff of smoke." This characteristic appearance was observed in angiography, which is used for diagnostic criteria.[2] It was first explained in 1957 titled as "hypoplasia of the bilateral internal carotid arteries." In 1969, Suzuki and Takaku given the descriptive title of "moyamoya."[3]

Moyamoya disease was previously considered to predominantly affect the Asian population.[4] But now, it affects people with different ethnicities, with progressive cases in American and European populations.[5] It is the most common neurovascular disorder among children in Japan with a prevalence rate of 3/100,000.[3] In Europe, the incidence of MMD is an about one-tenth of that in Japan and incidence of about 1 in a million in the United States.[3] The incidence rate was 4.6 for Asian Americans, 2.2 for African Americans, and 0.5 for Hispanics when compared with whites.[5] The disease is known to occur in both children and adults. It is two times more prevalent in females than males.[3]

Though genetic factors have an important role, the etiopathogenesis of MMD is clearly idiopathic.[3] The clinical signs and symptoms are headaches, convulsive seizures, silent ischemic attacks, involuntary movements, cognitive variations, and cerebral infarctions in the area of the frontal lobe. There may be progression of unilateral into bilateral lesions.[6]

The radiographic features include thinning of the terminal segments of the internal carotid arteries along with the development of collateral vessels.[7] The MRI reveals the characteristic feature of reduced flow in the cerebral branch of internal carotid arteries with prominent collateral flow voids in the basal ganglia and thalamus.[8,9]

The case report describes the specific clinical findings and dental treatment in a 12-year-old child presenting with MMD.

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Case Description

A 12-year-old male patient reported to the Department of Pedodontics and Preventive Dentistry with a chief complaint of broken tooth in the upper front tooth region due to trauma, 1 week back. The patient gave a history of prediagnosed case of MMD with hemiplegia of the right side.

At the age of 1½ years, the patient suffered from pulmonary tuberculosis and completed anti-tuberculosis treatment for the same. At the age of 4 years, the patient experienced sudden onset of pain and weakness in the right lower limb, which was associated with difficulty in holding objects with the right hand and raising the right arm above the head. The MRI of brain showed focal right frontal infarct with left cerebral atrophy and an attenuated appearance of intracanal arteries suggestive of effects of MMD. Symptomatic medical treatment was provided to the patient.

Fig. 1: General physical examination

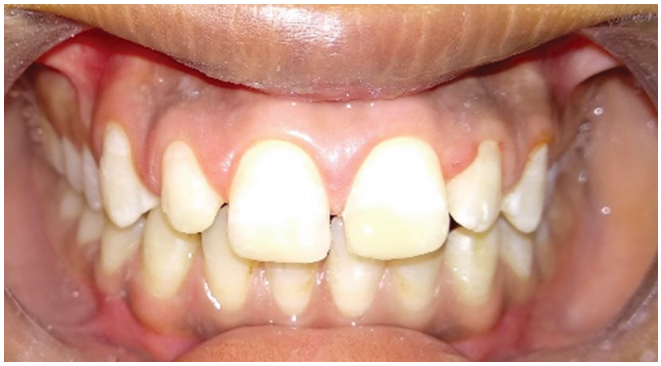

Fig. 3: Intraoral postoperative picture depicting composite restoration of upper central incisors

The patient had no past dental treatment exposure. There is no related family history.

The patient appeared underweight with a lean body type. The general physical examination revealed hemiplegia of the right side, affecting both upper and lower limbs (Fig. 1).

The extraoral examination revealed a symmetrical face with competent lips and normal mouth opening. On intraoral examination, permanent dentition was present with Angle's Class I malocclusion. Ellis Class I fracture was present in the upper right central incisor whereas Ellis Class II fracture was present in the upper left central incisor (Fig. 2).

The dental treatment plan focused on providing preventive as well as restorative care to the patient while simultaneously providing proactive counseling to both the parents and the patient. Behavior management was an integral part of our management. Oral hygiene measures were instructed to the patient and modified angled toothbrush with fluoridated toothpaste was prescribed due to the lack of manual dexterity. Along with this, diet modification was done by recommending intake of noncarious dietary food items such as fibrous food, nuts, and fruits. The medical consent was obtained from the pediatrician following which oral prophylaxis and composite restoration of upper central incisors were done (Fig. 3).

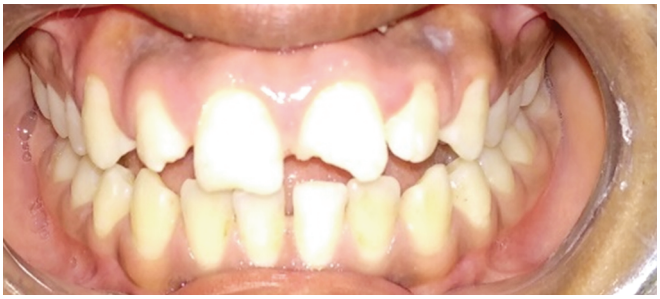

Fig. 2: Intraoral picture depicting fractured upper central incisors as chief complaint

Discussion

Moyamoya disease is a cerebrovascular disorder that involves narrowing of cerebral internal carotid artery and its branches. These moyamoya vessels are called collateral vessels that are formed in an attempt to supply blood to the brain after the occlusion of main cerebral arteries. They are very small, weak, and prone to bleeding, aneurysm, and thrombus formation.[10]

Asian population is mostly affected with MMD when compared with other ethnical background.[2] Giroud et al.,[11] Joarder and Chandy,[12] and Kim[13] reported cases of MMD that predominantly affected individuals of Asian heritage. In the present case, a 12-year-old male of Indian origin presented with clinical features of this rare disease.

An increased genetic predisposition is suggested from the fact that 6–10% of the patients have affected first-degree relatives.[14] Various gene loci such as 3q24-p26, 6q25, 8q23, and 17q25 have been identified in genome-wide analysis for familial MMD.[15] Multifactorial causes such as hepatitis-C virus-infected persons, cryoglobulinemia, sickle cell disease, and individuals after radiotherapy of tumors of the optic chiasm are also contributory to the etiology of the disease.[2] Therefore, the disease is considered to be both hereditary and multifactorial in nature. In the present case, no known cause was identified and the family history was inconclusive.

The clinical features of the present case included dysarthria, aphasia, stroke, and hemiparesis on the right side. This is in accordance to the cases reported by Goyal et al.,[16] Kim,[13] Okhovat and Moeini,[17] Janda et al.,[18] and Houba et al.[19]

The MRI seems to be more capable of representing morphological alterations of the central nervous system.[20] In the present case, MRI revealed focal right frontal infarct with left cerebral atrophy with attenuated appearance of intracanal arteries. Similar findings were reported by Knierim and Woodward,[20] Kim et al.,[21] and Yu et al.[22]

Several systemic diseases have been related with MMD.[23–25] The present case was associated with tuberculosis at the age of 1½ years.

A thorough knowledge about the clinical features and potential implications of MMD should be acquired to provide high-quality oral health care to the patients. As pediatric dentists, one should aim at providing holistic behavior counseling to these patients. The lack of manual dexterity in children with MMD leads to an inability to practice the various oral hygiene measures correctly. Hence, both the parents and the patients should be instructed about the various preventive strategies such as brushing, dietary alterations, and early identification of oral diseases. Therefore, it is imperative to maintain normocapnia, normotension, and effective pain control during dental procedures such as extraction to prevent postoperative ischemic complications due to reduced cerebral blood flow.

Conclusion and Case Importance

Moyamoya disease is a rare progressive cerebral arteriopathy associated with risk of ischemic and hemorrhagic brain complications. A thorough assessment of pertinent factors such as medical history and degree of involvement must be made prior to formulation of the treatment plan. Dental professionals must attain a complete knowledge of the various aspects of this anomaly so that an early diagnosis and monitoring from the initial stages can be undertaken to avoid any harmful implications. As pedodontists, one must be prepared to deal with the challenges that may arise during the diagnosis of such rare diseases and provision of effective treatment. The importance of counseling and reinforcement of preventive measures must be well catered. The goal should be provision of a treatment plan that can benefit the patient in the long run and help attain superior outcomes providing psychological relief to the patient at large.

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