Case Report
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A Child with Acute Hemiparesis: A Case Report

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Abstract

This is a case of an eight years old boy initially presented with right sided hemiparesis. His diagnosis was Moyamoya disease and been treated conservatively at initial stage. This disease is recurrent in nature, and the patient developed another stroke episode within 3 months. He was referred to neurosurgical team and was planned for a revascularization surgery for a better long-term outcome.

Keywords: Peadiatrics stroke; Moyamoya disease; Cerebrovascular accident; Stroke; Peadiatrics; Hemiparesis

Case Presentation

In pediatric age group, we rarely see a case of acute stroke as much as we see cases of adult stroke daily. There is a cerebrovascular disease in children of unknown etiology originate from Japan known as Moyamoya disease. We are reporting a case of eight years old boy with the complaint of right sided upper and lower limbs weakness to a hospital in Malaysia, and his diagnosis was Moyamoya disease. His stroke episode recurred in three months and the presentation worse than the first episode.

An eight-year-old Malay boy presented with sudden onset of right sided body weakness and numbness a day prior to hospital admission. There was no history of head injury, headache, and loss of consciousness or seizure. Patient did not show any behavioral changes or cognitive impairment after the development of weakness. On examination, the patient was fully conscious and well orientated. There was no dysmorphism, no neurocutaneous stigmatas, and his growth parameters are normal. He was afebrile with BP within 50th centile, pulse rate was 110 beat per minute and respiratory rate 26 per minute. Neurological examination showed no signs of meningeal irritation, with reduced power and tone over his right upper and lower limbs. No abnormalities seen on cardiorespiratory and abdominal examination.

A battery of blood tests including thrombophilia screening did not show any significant findings. CT brain features an acute infarction of left precentral and postcentral gyri. MRA showed obliteration of right MCA. Small and narrowing of the left MCA. Multiple collaterals are seen at the region of basal ganglia bilaterally, represent Moyamoya appearance. Patient was treated conservatively with aspirin 5mg/kg and was put under rehabilitation program. His hemiparesis was improved in 3 months when he able to walk as usual without support and he regained almost full muscle power.

After 3 months of the first episode, the patient developed another stroke attack. He had bilateral upper and lower limbs weakness. CT brain showed right parietal acute infarct with left frontoparietal cystic encephalomalacia with cortical laminar necrosis, no intracranial bleed. Cerebral angiogram revealed severe stenosis of ICA bilaterally with collaterals that are arising from PCA and PCOM supplying the high parietal and parasagittal segments. Patient developed hypertension and he was started with oral anti-hypertension. He was referred to neurosurgical team and currently a revascularization surgery was planned on him. As this case is written, patient's condition is improving. He able to get up from supine position and ambulating by rolling over his body. He was put under rehabilitation team for wheelchair ambulation and other muscle strengthening activities. The patient is awaiting his turn to undergo the operation.

Discussion

In Moyamoya disease, the cerebrovascular circulation is progressively occluded with neovascularization of arterial collaterals. This is a rare illness with annual incidence of 0.35 to 0.94 per 100,000 populations and the prevalence 3.2 to 10.5 per 100,000 populations. Ten to twelve percent of patients had a significant family history of Moyamoya disease. Although originally this disease considered affecting predominantly persons of Asian heritage, we can find the case reports

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throughout the globe in people of many ethnic backgrounds [1].

Moyamoya was derived from Japanese word for puffy and hazy, to describe the puff of smoke in the air. This term explained the appearance of the neovascularization of the collateral networks been visualized in angiogram as smoky and hazy like appearance. This was first reported in Japan since 1957 and subsequently there were much more similar cases revealed mainly in Japan and other part of Asian countries [2].

In children, the most common presentation is that of recurrent episodes of cerebral ischemia manifesting clinically as focal deficits, paresthesia, and seizures. The tapering of cerebral vessels was due to its respond to multiple external stimuli, injuries or genetic abnormalities. According to literatures, Moyamoya disease had found to be associated with thyroiditis, renal artery stenosis, antiphospholipid syndrome, congenital heart defects, Down syndrome, neurofibromatosis and sickle cell anemia. Once the vessel occlusion process started, it tends to continue further in spite of medical treatment given unless surgery is the management option [3].

Tapering and blocking of proximal vessels of cerebral flow with extensive formation of collateral through the new perforating vessels is seen as the classic appearance of puff of smoke in MR angiogram [4].

Symptomatic management in acute phase of the illness is important to improve cerebral blood flow, reduce the elevated intracranial pressure hence, and control the seizures. In patient with persistent neurological deficits rehabilitation is recommended to improve functional outcome [5].

Majority of the literatures recommend the revascularization surgery, as favorable outcome has been reported in terms of improvement of symptoms and positive angiographic follow ups in all age groups. Improves in hemodynamics post operatively seems to be similar in all age [6].

This case highlights a rare cause of acute pediatric stroke to be considered in differential diagnosis especially in Asian population. Early diagnosis and intervention are important to lead a better outcome in children with Moyamoya disease.

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