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Case Study

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MOYAMOYA DISEASE: CASE REPORT

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ABSTRACT

Moyamoya disease is a condition characterized by progressive narrowing of Intracranial arteries around Circle of Willis and development of collaterals. This case report describes a 12-year-old Ukrainian Girl, migrated to Ireland couple of months ago, presented with right sided facial palsy and right hemiparesis of sudden onset. Her MRI brain revealed acute infarct in MCA territory with loss of flow in both proximal and middle cerebral arteries. CT Angiogram showed abnormal appearance of ICA and circle of Willis. progressive narrowing of right MCA and high-grade narrowing of left MCA suggestive of Moyamoya disease. Conservative management plan was decided Prednisolone, aspirin in tapering dose was started, she showed complete recovery in couple of weeks. There was no underlying association with any haematological, immunological or infectious cause was identified and there was no family history of such event. Moyamoya disease has mortality rate of 4.5% is children and majority of children develop gradual cognitive decline over time.

INTRODUCTION

Moyamoya disease is a progressive occlusion of the cerebral vasculature.[1,2] Moyamoya disease has the highest incidence among certain populations like Asian and Japanese. The male-to-female ratio is approximately 1:1.8. Most commonly Moyamoya is diagnosed during the first decade of life[2,5] Typically, the occlusive process affects both sides of cerebral blood vessels, although there have been documented cases of unilateral involvement.[3,4] Here, we report the case of a patient with Moyamoya disease who presented with stroke involving right side of body.

CASE PRESENTATION

A 7-year-old Girl weighing 37 kg presented with sudden onset of left sided body and facial weakness. There was no associated history of loss of consciousness, seizures, head trauma, vomiting or bladder-bowel incontinence. Her weakness on arm and leg recovered quickly in couple of hours but facial weakness persisted.

She was born to nonconsanguineous parents as normal vaginal delivery in Ukraine and family moved to Ireland about 2 years ago. She was attending normal stream school and had no learning difficulties. During the physical examination, her vital signs and anthropometric measurements were within the normal range. Muscle power on left was 4 out of 5, tendon reflexes were brisk, gait was normal except in turning of left foot while walking. She had left sided facial palsy. Systemic examination remained normal. Laboratory investigations revealed a normal blood count, coagulation profile, Renal and liver functions, Her thyroid function, Compliment levels, inflammatory markers, Thrombophilia screen, Hb Electrophoresis, plasma Homocysteine levels, Varicella zoster titres, ANA, ANCA and Lupus anti-coagulant were all normal. CSF examination was unremarkable and PCR and culture remained negative. Chest X-ray, ECG and echocardiography revealed no abnormality. MRI of the brain and CT Angiogram was suggestive of acute infarct in right Middle cerebral territory with loss of flow in both proximal and middle cerebral arteries.

Patient was commenced on IV methyl Prednisolone for 5 days and Aspirin was also started, had input from physiotherapy, SLT and occupational therapy.

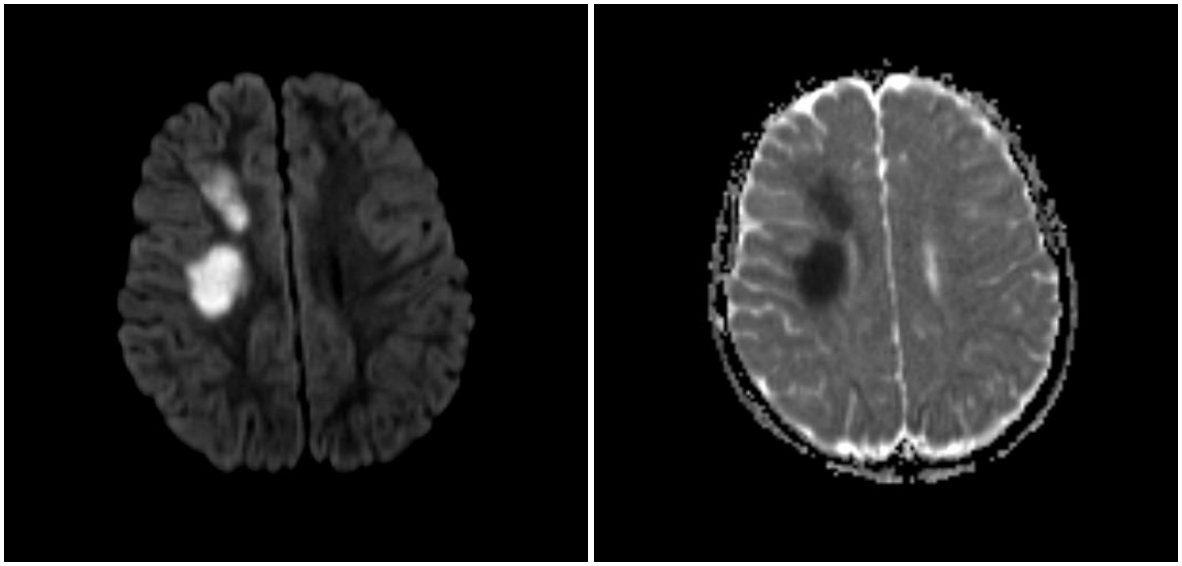

Figure 1: MRI image demonstrating increased b1000 diffusion signal (top) and diffusion restriction (bottom) in the right frontal centrum semiovale indicating an acute infarct (in the right middle cerebral artery territory.

Figure 2: Maximum intensity projection image from the CT angiogram which demonstrates the superior part of the right internal carotid artery (hollow arrow) which discontinues abruptly, and the expected location of the right middle cerebral artery (block arrow).

DISCUSSION

Moyamoya Disease was named in 1969 by Suzuki and Takaku[2] because the appearance on angiography is reminiscent of a "puff of smoke" (the Japanese word moyamoya is translated as "something hazy like a puff of smoke") due to the formation of the collateral vessels. Moyamoya disease is a progressive disease-causing occlusion of vessels of circle of Willis.[3] The occlusive process initially starts from intracranial portion of internal carotid arteries and progressively involves middle, anterior and posterior cerebral arteries and development of characteristic collateral vessels.[4] Angiographic appearance of these collateral vessels gives appearance of something hazy like Puff of cigarette smoke, that gives it name Moyamoya, a Japanese word meaning puffy.

Moyamoya Disease (MMD) is occurrence of cerebral Vaso occlusive phenomenon without underlying medical condition. **Moyamoya syndrome** (MMS) is characterized by moyamoya angiographic findings due to underlying medical conditions.[7] MMD has been associate with Grave's disease, tuberculosis, and hematologic disorders such as Fanconi anaemia, sickle cell anaemia, Hereditary spherocytosis, PK Deficiency, protein S deficiency and lupus anticoagulant. Additionally, coarctation of the aorta, fibromuscular dysplasia, Renal artery stenosis, cranial trauma, radiation injury and hypertension have been linked to the disease.

The symptoms of the disease can vary widely from being asymptomatic to presenting with severe neurological deficits.[13] It presents in children (2/3rdof cases) or in adulthood around 30-40 years of age.[7] In children, ischemic stroke is the most common presentation, In adult most commonly Moyamoya presents as TIA or haemorrhagic stroke. It may also present with headache, seizures and other neurological symptoms. Cognitive impairment can occur due to chronic cerebral hypoperfusion and recurrent ischemic insults.[5,6,8]

Cerebral infarct often occurs in cortical and subcortical regions distal to occluded moyamoya vessel, Intra cranial Haemorrhage occurs in deeper structures such as basal ganglia, thalamus or ventricular system. CTA or MRA are non-invasive imaging can demonstrate characteristic findings of Moyamoya. Digital subtraction angiography (DSA) is a gold standard for diagnosis of Moyamoya. DSA is an invasive procedure and involves arterial puncture and has risk of infection, haematoma at puncture site, risk of arterial dissection and stroke.[10]

In Acute stage treatment of Moyamoya disease is symptomatic and goal is to maintain cerebrovascular perfusion. Anti platelets are used is acute cases and as prevention of further strokes.[7] Corticosteroids are not primary treatment of Moyamoya disease and is considered as standard therapy where moyamoya disease is associated with vasculitis.[10]

There is no curative treatment of Moyamoya, supportive management can reduce the risk of complications, surveillance by imaging can identify patients at risk of ischemic or haemorrhagic stroke and can be benefitted from surgical revascularization.[12,13,14,15]

Moyamoya is a progressive disease, 50-66% of patients have poor outcome, often with cognitive decline and other neurological complications.[2,3,16]

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