Case Report

Moyamoya disease in a case of tuberculous meningoencephalitis

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ABSTRACT

Moyamoya disease is an uncommon vascular disease, which causes obstruction and stenosis of arteries of the circle of Willis. This disease is seen across the world, but is more common in east Asia. It may cause hemorrhagic or ischemic stroke, or transient ischemic attack. Fever, vomiting and convulsions, are usually suggestive of infective etiology involving brain and meninges. This case report is about a child with meningoencephalitis. Tuberculous meningoencephalitis is the commonest cause, in India, followed by viral type. In our patient we found out that there was an additional finding in form of moyamoya arteritis, which is a rare finding in children.

Keywords: Moyamoya disease, Meningoencephalitis, Arteritis

INTRODUCTION

Moyamoya disease is a rare vascular disease, progressive in nature, in which arteries of the circle of Willis becomes obstructed or stenosed, reducing blood flow to brain.¹⁻³ Moyamoya disease is more common in east Asian countries as compared to the rest of the world. There are two peaks of onset in moyamoya disease one is in childhood and the other is in young adulthood. Moyamoya disease may cause transient ischemic attack (tia), stroke, hemiplegia which are recurrent with aneurysm, or bleeding in the brain.²⁻⁶

CASE REPORT

A 4-year-old child was brought to the hospital with history of fever in the last 7 days. The child was drowsy and had generalised convulsions when the child was brought to us at Bharat Ratna Dr. Babasaheb Ambedkar municipal general hospital, Mumbai, India. The child was drowsy disoriented and having persistent convulsions. The convulsions were controlled with anti-convulsant and a lumbar puncture was planned. An ophthalmology examination showed that the child was having

papilledema, suggestive of raised intra cranial tension. Hence, the lumbar puncture was deferred. The reflexes of the child were brisk and there were signs of raised intra cranial tension, also signs suggestive of meningitis were present like neck stiffness and irritability with projectile vomiting. In view of the above history and examination findings a provisional diagnosis of meningoencephalitis was thought off. The commonest cause is either tuberculosis or viral meningitis for the about presentation. Anti-oedema medications were given and a guarded lumbar puncture was done which was suggestive of tuberculosis, even after three to four days of medications, the child did not show any improvement. A MRI brain was done which showed multiple patchy wedge shaped areas of restricted diffusion in right anterior cerebral artery territories involving cortical and subcortical regions of basi-frontal and fronto-temporo-parietal lobes, right thalamus, bilateral basal ganglia and left hippocampal regions suggestive of acute infarcts with critical stenosis involving supra- clinoid- cavernous ICA, a1 segment ACAs and m1 segment of MCA, generalised irregularity with variable degree of luminal narrowing involving intracranial arteries, suggestive of moyamoya disease. This was confirmed by DSA correlation. Also,

abnormal lepto-meningeal enhancement was seen along the bilateral cerebral convexities confirming the meningoencephalitis. Specific tests for hypercoagulability such as APLA, ANA antibodies, basal homocysteine levels were also assessed, the results for which were normal. An EEG was suggestive of bilateral parietal occipital slowing. Summarizing all this we have a child with a rare arteritis with a tuberculous meningoencephalitis like picture. The child was started on anti-Koch treatment and anticonvulsants. Neurosurgery was not done, but low dose anti coagulants were started. Child improved and was discharged but residual hemiparesis was present with slurred speech. Normally moyamoya disease presents with recurrent hemiparesis but in our case, we had a meningoencephalitis picture with a combination of tuberculosis and an unexpected arteritis. Anticoagulants were started in addition to anti Koch's treatment and the child improved clinically.

Figure 1: Puff like appearance in moyamoya arteritis.

Figure 2: MRI angiography image of the child.

DISCUSSION

Moyamoya disease is a rare, progressive vascular disorder in which arteries of the circle of Willis become obstructed or stenosed, reducing the blood flow to the brain.¹⁻³ Tiny blood vessels then develop at the base of the brain in an attempt to supply blood to the brain. The word “moyamoya” means “puff of smoke” in Japanese, a term describing the appearance of net-like tiny blood vessels. Moyamoya disease is found all over the world, but it is more common in east Asian countries, especially Japan, Korea, and China, and in people of east Asian descent.⁴ There are two peaks of onset in moyamoya disease; one is in childhood, and the other is in young adults. Ischemic stroke is dominant in children and hemorrhagic stroke is dominant in adults.²⁻⁶ In autopsy, intimal hyperplasia, atrophic media, and widening of the internal elastic lamina can be seen in affected arteries.^{2,5} Moyamoya disease may cause TIA, stroke, aneurysm, or bleeding in the brain.^{2,5,6} It can also cause cognitive and developmental delays or disability. The confirmatory tool to diagnose the disease after MRI is magnetic resonance angiography (MRA). It reveals narrowing and occlusion of proximal cerebral vessels with blood flowing through collaterals giving puff of smoke appearance. Cerebral angiography of moyamoya disease shows stenosis and occlusion of arteries of the circle of Willis with moyamoya vasculature as developed collateral circulation.^{2,3,7-9} Flow void sign suggests moyamoya vasculature on MRI in bilateral basal ganglia.⁷⁻⁹ Electroencephalography shows rebuild up of delta waves once hyperventilation is stopped, which is a typical finding in moyamoya disease.^{2,10} Medications are prescribed to reduce the risk of stroke or to control seizure.² Aspirin or other antiplatelet drugs are recommended to prevent strokes. Calcium channel blockers may relieve headaches. Anti-seizure medications should be prescribed if the patient has seizure disorder. If symptoms become worse or if tests show evidence of low blood flow, revascularization surgery is recommended. Direct or indirect revascularization procedures, or a combination of both may be used. Indirect procedures include encephaloduroarteriosynangiosis or encephalomyosynangiosis.²

CONCLUSION

A rare arteritis like moyamoya disease could be present additionally in other forms of meningoencephalitis.

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