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CASE REPORT

A Combination of Moyamoya Pattern and Cerebral Venous Sinus Thrombosis: A Case of Tubercular Vasculopathy

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ABSTRACT

Background: Moyamoya vasculopathy, arising secondary to tubercular meningitis (TBM) is unusual. There have also been a few reports of cerebral venous sinus thrombosis (CVST) in TBM. A case of TBM, complicated simultaneously by Moyamoya syndrome and CVST, is being presented here.

Case: A 1-year-old girl presented with febrile encephalopathy, vomiting, seizures and left hemiparesis. Cerebrospinal fluid analysis was suggestive of TBM. Extensive infarcts were noted in the magnetic resonance imaging, involving right middle cerebral artery (MCA), anterior cerebral artery and the left MCA. Magnetic resonance venogram revealed left transverse venous sinus thrombosis and magnetic resonance angiography showed bilateral moyamoya pattern of arteriopathy. Patient was started on antitubercular therapy and low molecular weight heparin.

Conclusions: Early vascular involvement affecting both arterial and venous structures has not hitherto been reported in CNS tuberculosis. Early recognition of secondary complications of CNS tuberculosis is crucial to prevent the morbidity and mortality associated with TBM.

KEYWORDS: tubercular meningitis, moyamoya pattern, cerebral venous sinus thrombosis, tubercular vasculopathy.

INTRODUCTION

Prevalence of tuberculosis (TB) is high in developing countries with estimated worldwide prevalence of 13.7 million (206 per 100 000 population) in 2007 [1]. Around 10% of these cases develop tubercular meningitis (TBM). Neurotuberculosis may manifest in the form of meningitis, meningoencephalitis, tuberculoma, tubercular abscess or stroke owing to tuberculous vasculitis [2]. Early recognition of the secondary complications is crucial in limiting the associated morbidity and mortality. Moyamoya pattern of vasculopathy secondary to infectious stimuli, such as bacterial meningitis, is a well-known entity, whereas TBM complicated by

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moyamoya pattern of vasculopathy is exceedingly rare [3]. There have also been a few case reports of TBM predisposing to cerebral venous sinus thrombosis (CVST) [4]. A case of a 1-year-old girl with TBM complicated by both moyamoya pattern of vasculopathy and CVST simultaneously is being presented here. To the best of our knowledge, there has been no previous report of TBM complicated by both Moyamoya syndrome and CVST simultaneously.

CASE SUMMARY

A 1-year-old girl presented with low-grade, intermittent, daily fever for past 1 month, vomiting and altered sensorium for past 8 days, paucity of movements in the right upper and lower limbs for past 8 days preceded by an episode of generalized convulsion. There was no apparent cranial neuropathy, flaccidity or tightness of limbs, other involuntary movements, jerky eye movements, sensory complaints or bowel bladder disturbances. There was no evidence of prolonged cough, cachectic symptoms and contact with a case of TB. There was no preceding history of trauma, prodromal illness, vaccination or animal bite. There was no history suggestive of joint pain, swelling, bloody urine, previous blood transfusion or any cardiac illness and abnormal odor of body fluids.

On examination, her vitals were normal and fundus examination did not reveal any papilloedema. Salient findings on neurological examination were a comatose child with right hemiparesis, generalized hyperreflexia and neck rigidity. Rest of the neurological and systemic examinations was normal.

The possibility of a subacute febrile encephalopathy with vasculitis, secondary to TBM or complicated bacterial meningitis, was kept. The child was started on intravenous antibiotics (Ceftriaxone and Vancomycin). She required mechanical ventilation for 3 days in view of poor sensorium. During hospital stay, she also had features of raised intracranial pressure (ICP) in the form of intermittent posturing, which responded to medical measures.

Her hematological investigation revealed anemia (hemoglobin level of 7.4 g/dl), with the peripheral smear showing microcytic hypochromic anemia with increased red cell distribution width suggesting

iron-deficiency anemia. Her cerebrospinal fluid (CSF) study showed 400 cells (60% lymphocytes, 40% polymorphs) with elevated protein (540 mg%) and hypoglycorrhachia (CSF sugar 30 mg/dl against a random blood sugar of 86 mg/dl). Magnetic resonance imaging (MRI) brain (Fig. 1A–D) showed infarcts extending over right middle cerebral artery (MCA), anterior cerebral artery and left MCA territories with mild ventriculomegaly. There were no ring-enhancing lesions or meningeal enhancement. Magnetic resonance angiography (MRA) (Fig. 2A) showed bilateral moyamoya pattern of arteriopathy with distal internal cerebral artery and proximal MCA narrowing without any collateral formation. Magnetic resonance venography (MRV) (Fig. 2B and C) showed thrombosis of the left transverse sinus. A diagnosis of complicated TBM was made and the child was started on antitubercular drug therapy with steroids and low molecular weight heparin.

The child responded to these measures within 3–4 days and was extubated. There was rapid improvement in her sensorium, and resolution of the features of raised ICP.

Antinuclear antibodies and antiphospholipid antibodies were not detected. Procoagulant work up in the form of serum antithrombin III, protein C, protein S, β_2 glycoprotein and factor V levels done after 12 weeks of the event did not reveal any abnormality.

DISCUSSION

CNS TB may manifest as meningitis, meningoencephalitis, tuberculoma, tubercular abscess, or stroke owing to tuberculous vasculitis. One of the most severe complications of TBM is vasculitis in the vessels of the circle of Willis, the vertebrobasilar system, and the branches of the MCA. Rich and McCordock attributed vasculitis to a hypersensitivity response [5].

Rarely, tubercular vasculitis causing moyamoya pattern of vasculopathy has been described. It has been reported as a parainfectious vascular phenomenon, complicating either bacterial or TBM, usually occurring within 2 weeks of disease onset [6]. In a case reported by Umesh K [7], a girl child who had TBM at the age of 1 year, later on presented with a seizure at the age of 10 years, which was attributed to Moyamoya syndrome.

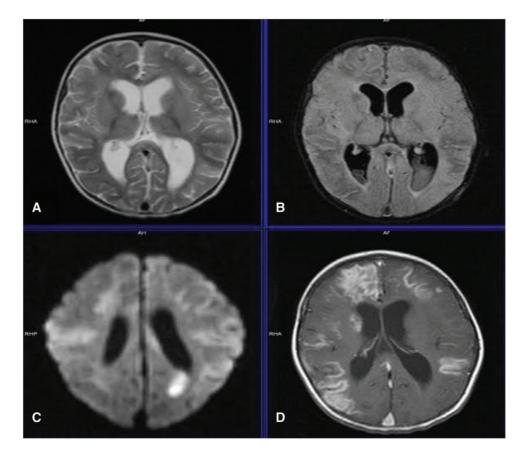


Fig. 1. T2 weighted (A) and FLAIR (B) axial MRI brain images show hydrocephalus with subtle signal changes in bilateral cortical gray matter. Diffusion weighted image (C) shows restricted diffusion in the bilateral gray matter and left posterior periventricular white matter. Contrast enhanced T1 weighted axial image (D) shows gyral enhancement in cortical gray matter and right basal ganglia, consistent with ischemic infarcts.

TBM can rarely predispose to CVST [4]. Multiple pathophysiologic mechanisms have been proposed including injury to the endothelium by inflammation, increased platelet aggregability and alterations in blood flow [8]. In a study by Mangalore et al. [9], imaging and pathological features of two autopsy-proven cases of tuberculous thrombophlebitis were described, with venous infarcts involving the superficial venous system in one and the deep venous system in another. They proposed that tuberculous thrombophlebitis should be suspected if basal exudates and multiple white matter T2 hyperintensities are seen on neuroimaging. In the current case, although there were no basal exudates, but presence of periventricular white matter signal changes, encephalopathy and features of raised ICP prompted the evaluation for venous sinus thrombosis.

In the current case, the patient had iron-deficiency anemia, which could have contributed both to arterial and venous vasculopathy.

Moyamoya syndrome typically develops within 2 weeks of TBM. Early detection of these vascular complications is of paramount importance. Treatment of tubercular vasculitis is essentially antitubercular therapy and addition of steroids, whereas presence of CVST would necessitate initiation of anticoagulation therapy with unfractionated or low molecular weight heparin.

This case report highlights that in a case of bacterial or TBM, presence of focal neurological deficits, encephalopathy and raised ICP (out of proportion to the degree of hydrocephalus) should prompt one to rule out underlying vasculopathies, both arterial and venous, as this has both immediate

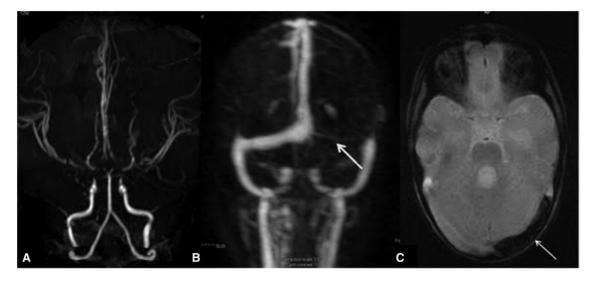


Fig. 2. MRA (A) shows occlusion of bilateral supraclinoid internal carotid artery with reformed but attenuated anterior cerebral and MCA, consistent with a Moyamoya pattern. MRV (B) shows non-visualization of the left transverse sinus, which shows the presence of a thrombus on gradient T2 axial image (C).

therapeutic and long-term outcome-related implications.

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