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MOYAMOYA DISEASE PRESENTING AS ACUTE ENCEPHALITIS SYNDROME

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ABSTRACT

Background: Moyamoya disease (MMD) is a chronic, progressive occlusion of the circle of Willis arteries usually affects bilateral carotid arteries than unilateral that leads to the development of characteristic collateral vessels seen on imaging, particularly cerebral angiography. Once the occlusion process begins, tends to continue despite any known medical management unless treated with revascularization surgery

Report of the case: A 28 year female gravida four in 34th week of gestation presented with features suggestive of acute meningoencephalitis and CSF examination suggested 75% polymorphs of 300 total leukocytes. After 7 days of optimum management, complaints of headache aggravated so patient was subjected to MRI Brain which reported as early subacute intraparenchymal haemorrhage left frontal lobe in periventricular region with mild surrounding perifocal edema and intraventricular extension. On further MR angiogram, patient was diagnosed as MMD. Patient was asymptomatic in her previous 3 pregnancies and presently underwent planned preterm high risk caesarean section after a week. However condition of patient deteriorated over 2 weeks post delivery, did not remain fit for any neurosurgical intervention and unfortunately succumbed to death.

Conclusion: Antenatal patient presenting as acute meningoencephalitis with persistent headache refractory to medical therapy, should be subjected to CT/MR brain imaging for rare possibility of MMD and early neurosurgical interventions in view of its grave prognosis.

KEYWORDS : Moyamoya Disease, Meningoencephalitis, Pregnancy

INTRODUCTION

Moyamoya disease (MMD) is a chronic, progressive stenosis of the circle of Willis arteries affecting the distal intracranial carotid arteries and their proximal branches that leads to the development of characteristic collateral vessels visible on cerebral angiography. The disease may develop in children and adults, but the diagnosis can be challenging given the spectrum of overlap between various other arteriopathies.

CASE REPORT

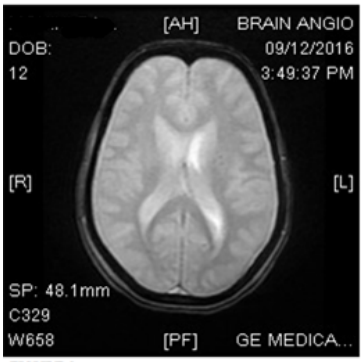
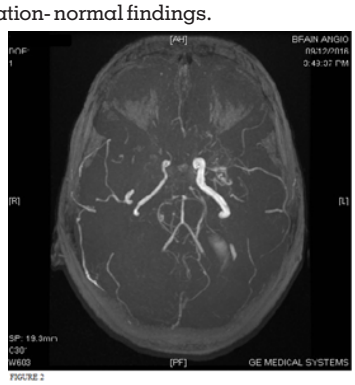


FIGURE 1

This 28 year old, 34 weeks antenatal mother with G4P3L3A0 obstetric history was admitted with the chief complaints of headache, vomiting and fever since 3 days. Patient was apparently alright 3 days back when she developed sudden onset bitemporal headache in afternoon which was constant in nature and associated with altered level of consciousness without any aggravating and relieving factors. The headache was associated with 3 episodes of vomiting associated with nausea, non projectile non bilious watery with food contents in nature not associated with any hemetemesis. There was history of high grade fever associated with chills and rigors. No history of loss of consciousness hemiparesis, vertigo, tinnitus, seizures, abnormal body movements. History of mother died 2 years back due to some similar complaints of severe headache of which no records could not be retrieved. No other relevant history was present. General condition below average, afebrile, pulse 80bpm, respiration 16/min thoracoabdominal, b.p 100/60 mmHg gait ataxia present. Cranial nerve examination are all normal. CNS examination - patient is conscious oriented to time place person. Nutrition of limbs are B/L normal, reflexes are normal. Signs of meningeal irritation were present neck rigidity present kernigs sign positive. Pupils are bilaterally normal and reacting to light. Planters on right side flexors and left side extensors. RS CVS P/A examination- normal findings.



On investigation patients hemogram was suggestive of acute neutrophilic infective etiology while renal and liver profile were within normal range. Ultrasound obstetrics and abdomen suggested of normal upper abdomen study with single live fetus of 34 weeks with intrauterine cardiac activity with cephalic presentation with anterior placenta. Patient was subjected to CSF analysis with total leukocyte count 300 cells/mm^3, 75% polymorphs, RBC 80-100/hpf, proteins 31g/dl and sugar 28g/dl. Hence on the basis of clinical profile and laboratory elements the diagnosis of acute meningoencephalitis was made and patient was treated on line of acute encephalitis syndrome. Patient CSF sample was also sent for ADA levels, TBPCR, HSV-1, Japanese encephalitis and in coming days the all the reports were negative. After 10 days of treatment, patient improved clinically with resolving meningeal signs and GCS and MMSE score. Repeat CSF analysis was done suggestive of TLC 30-35 cells/mm^3, polymorphs 30%, RBC 0/hpf, proteins 28g/dl and sugar 66mg/dl. However with

clinical improvement of all the symptoms patient's headache persisted and gradually aggravated in last 3 days of 10 days hospitalization.

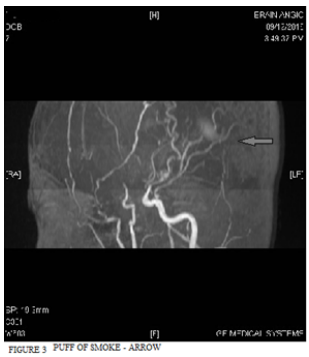


FIGURE 3 PUFF OF SMOKE - ARROW

Hence patient was subjected to MRI Brain which suggested of Early subacute intraparenchymal haemorrhage left frontal lobe in periventricular region with mild surrounding perifocal edema and intraventricular extension, multiple small altered signal intensities in bilateral frontoparietal White matter likely ischemic foci. I/v/o MRI findings suggestive of hemorrhage patient was henceforth subjected to MR angiogram which suggested of Severe diffuse narrowing in supraclinoid portions of bilateral ICA with near complete loss of flow related enhancement in bilateral MCA and ACA, Mild diffuse narrowing also noted in petrous and cavernous segments of right ICA. Diffuse narrowing also noted in basilar and bilateral PCA, Multiple collaterals channels noted in basal cistern and bilateral basal ganglia giving puff of smoke appearance with partial reformation of distal ACA, Left MCA and bilateral PCA. Left MCA has 3mm outpouring s/o small aneurysm . All findings s/o Moyamoya disease. Patient was planned for neurosurgical intervention after elective caesarean section. But patient condition deteriorated for next 2 weeks and was taken for emergency caesarean section but 2 days post delivery patient GCS worsened to 3/15 and was unfit for further neurosurgical intervention. Unfortunately patient could not be salvaged.

DISCUSSION

Moyamoya disease is an uncommon cause of stroke features progressive stenosis of internal carotid artery and formation of a collateral network of basal perforators (1). Being a chronic progressive occlusive condition it causes stenosis of intracranial internal carotid arteries and their proximal branches causing reduced blood supply to the anterior surface of brain, thereby leading to the formation of collaterals near the apex of carotids mimicking "Puffs of Smoke" known as Moyamoya in Japanese (2). The process of narrowing of cerebral vessels probably a reaction of cerebral blood vessels to a various range of external stimuli, injuries, or genetic defects (3). Originally considered to affect predominantly Asian heritage has now been observed round the globe. The incidence has bimodal peak age groups: children near 5 years old and adults in their mid 40's (4-7) with male-to-female ratio 1:1.8 or 1:2.2 in these surveys, and approximately 10%-15% had a family history. MMD risk in family members is around 30-40 times higher than the general population (4,5,8). Associations of MMD such as sickle cell anemia, neurofibromatosis-1, Down syndrome, congenital heart defects, antiphospholipid syndrome, renal artery stenosis, and thyroiditis have been found in the literature (3).

MMD usually presents with recurrent headaches and is migrainous in nature. Other presentations include TIA, ischemic stroke, hemorrhagic stroke, seizures, headache, and cognitive impairment and incidence of each symptom varies with the age of the patient(9). Stenosis once begins, tends to continue despite any known medical management unless treated with surgery (10). The early presentation of headache and vasculopathy gives us the differential diagnosis of PACNS, RCVS, and MMD. Features including recurrent thunderclap headache, noninflammatory CSF, presence of cortical subarachnoid hemorrhage, and sausage string appearance on angiography differentiates RCVS from vasculitis (11).

Acute management is primarily symptomatic and directed towards reducing raised intracranial pressure, improving cerebral blood flow, and controlling seizures (7).

Revascularization procedures are mainstay to improve the perfusion of the hypoxic brain tissue. Two main surgical revascularization procedures described: direct and indirect. Direct revascularization techniques, typically used in adults, include the superficial temporal artery to middle cerebral artery bypass or the middle meningeal artery to middle cerebral artery bypass (12-15). The Indirect surgical procedures, primarily used in children, aims to increase the volume of circulation in the collateral vasculature. This process requires neovascularization from the extracranial soft tissue to the poorly perfused cerebral areas and occurs within 3 to 6 months (16). Techniques include encephaloduroarteriomyosynangiosis,encephaloduroarterio synangiosis, encephalomyoarteriosynangiosis, and encephalomyosynangiosis (2,17,18). These procedures directed towards increasing perfusion in the cerebrovascular territory of the middle cerebral artery have no substantial effect in the anterior or posterior cerebral arteries territories. To manage the hemorrhagic and ischemic consequences of moyamoya disease surgical modalities are used (12,13,17,18,19,20,21) including ventricular drainage and hematoma evacuation for hemorrhagic cases (22). For ischemic MMD, surgical methods aims to restore and maintain adequate cerebral perfusion (12,13,17,18,,21,23). Long-term favourable outcome in terms of improvement in symptoms and positive angiographic follow-ups in all age groups has been reported in the literature for these procedures (7).

MMD has rapid and worse prognosis in children younger than 3 years compared to those aged 3 years or older (24). However, prognosis is generally worse for adults compared to children since adults have increased hemorrhagic episodes and henceforth higher mortality (16,25). To prevent additional stroke events and improve outcomes, careful, long-term neurologic and radiologic follow-up is required in adults with MMD.

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