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CASE STUDY

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Diagnostic dilemma of cognitive decline in a young female: considering congenital rubella, depressive disorder, and moyamoya disease

Abstract

We report a case of a 20-year-old young female patient from North India presenting with headache, decline in scholastic performance, irritability, and low mood, and having a history of hemiparesis, who had rubella infection in her antenatal period, who was investigated and diagnosed as persistent depressive disorder (PDD) with left-sided hemiparesis in remission with moyamoya disease (MMD) based on history, examination, and relevant investigation (magnetic resonance imaging [MRI] brain/magnetic resonance [MR] angiogram). We highlight the difficulties we faced in attributing whether the cognitive decline was due to congenital rubella, PDD, MMD, or a combination of any of these three diagnoses.

Keywords: Paresis. Rubella. Comorbidity. India.

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INTRODUCTION

Moyamoya disease (MMD) is a chronic progressive occlusive disease of the circle of Willis arteries that leads to the formation of the collateral vasculature at the base of the brain and is often an important predisposing cause of stroke in young age.[1] Terms such as "moyamoya syndrome" and "angiographic moyamoya" are often used when the disease is associated with comorbid conditions.[2] MMD is a rare disorder with prevalence of 0.086/100,000 population in the United States of America which was significantly lower than the 0.35 reported in the nationwide Japanese survey.[3] In India, prevalence data is not available.

Congenital rubella syndrome consists of sensorineural hearing defects, eye abnormalities such as cataracts, microphthalmia, retinopathy along with heart defects - the most encountered one being patent ductus arteriosus.[4] It rarely also affects the brain and presents as progressive rubella panencephalitis (PRP) or subacute sclerosing panencephalitis (SSPE).

We are presenting a case of a young female presenting with progressive cognitive decline and loss of interest in daily activities for four years. She had suffered hemiparesis four years back and was operated for congenital cataract.

CASE SUMMARY

A 20-year-old female was brought by her parents with the complaints of headache and decline in scholastic performance for past two and half years. The complaints were persistent and progressive in nature. She was having difficulty in academic performance with difficulty in understanding basic concepts of her syllabus with frequent forgetfulness and decreased attention span, difficulty in planning things and taking decisions following which she underperformed in her annual examinations of college. She also had episodic dull headache over the frontotemporal area with each episode lasting for two to three hours, which was mild in severity aggravated while studying and used to subside on itself. Frequency of headache was initially two to three episodes per week but for last three months was occurring daily with one to two episodes per day. Headache was not associated with any other symptoms. For last two years her social interaction had also decreased and most of the time she was withdrawn, spending her time alone

in her room with low mood and irritability most of the time of day. She had difficulty in initiation and maintenance of sleep with waking up early in the morning. Her intake of food had also reduced during this period.

Past history revealed that she developed sudden onset of weakness of the left upper limb four years back that progressed to involve the left lower limb within next two to three days. She also developed difficulty in swallowing solid food with difficulty in speaking and deviation of angle of mouth to the right side. She had an episode of confused behaviour with restlessness, irritability, and inability to recognise family members within few days of onset of the above symptoms. She underwent various investigations at the time of hemiparesis, records of which were not available. However, report of computed tomography (CT) scan of brain showed an infarct in territory of right middle cerebral artery. She was given tablet levetiracetam 1000 mg/day in two divided doses, tablet aspirin 75 mg/day, tablet clopidogrel 75 mg/day, and tablet atorvastatin 10 mg/day.

These symptoms improved over next one month; however, she had to continue physiotherapy as she had difficulty in maintaining posture and walking independently. She started walking after five to six months with support and after one and half years she became independently functional with minimal disability. Patient was diagnosed as having major depressive disorder (MDD) two year back and was given tablet escitalopram 10 mg with tablet clonazepam 0.5 after which there was improvement in her symptoms but was still having impairment in her scholastic performance. She left medication four months back on her own. Even though patient had improvement in mood, sleep, and social interaction still she had difficulty in cognitive and executive functioning with impaired scholastic performance while on medication.

During her antenatal period in first trimester her mother suffered rubella infection that improved within four to five days. Patient was delivered in hospital at full-term through normal vaginal delivery. She had delayed cry at birth and was weighing 1.2 kg. After three to four months postpartum, she was diagnosed with congenital cataract in left eye, for which she was operated. She had delayed motor and speech milestones, but her academic performance and social interaction was average before development of hemiparesis. At the age of four years she was diagnosed as having sensorineural hearing loss in the left ear for which she was advised to use hearing aid after the initial medical therapy.

At present she has low vision in her left eye (finger counting) and normal vision in right eye and decreased hearing from the left ear. Other general physical examination did not show any abnormality. Neurological examination revealed increased tone and decreased power (4/5) in the left upper and left lower limbs. Sensory examination of the patient was normal. Superficial reflexes were decreased on the left side of the body and deep tendon reflexes were brisk on the left side. Mental state examination of the patient revealed decreased psychomotor activity with low mood and restricted range of affect with pessimistic thoughts of hopelessness and worthlessness with cognitive examination showing poor concentration, impaired immediate and recent memory, and impairment in abstract thinking. Detail cognitive and executive assessment could not be performed as the patient was partially cooperative and got irritated during the examination.

Considering other medical issues, she was subjected to laboratory investigations. Her complete haemogram, liver function test, and kidney function test were within normal limits. Magnetic resonance imaging (MRI) brain revealed large ill-defined area in the right fronto-temporo-parietal area causing dilatation of the ipsilateral lateral ventricle (Figure 1). T2 weighted images showed hyper-intensities on the periventricular white matter and bilateral centrium semi-ovale which were isotense on the T1 weighted and hypertense on fluid-attenuated inversion recovery (FLAIR) images. Magnetic resonance (MR) angiogram showed thinned supraclined

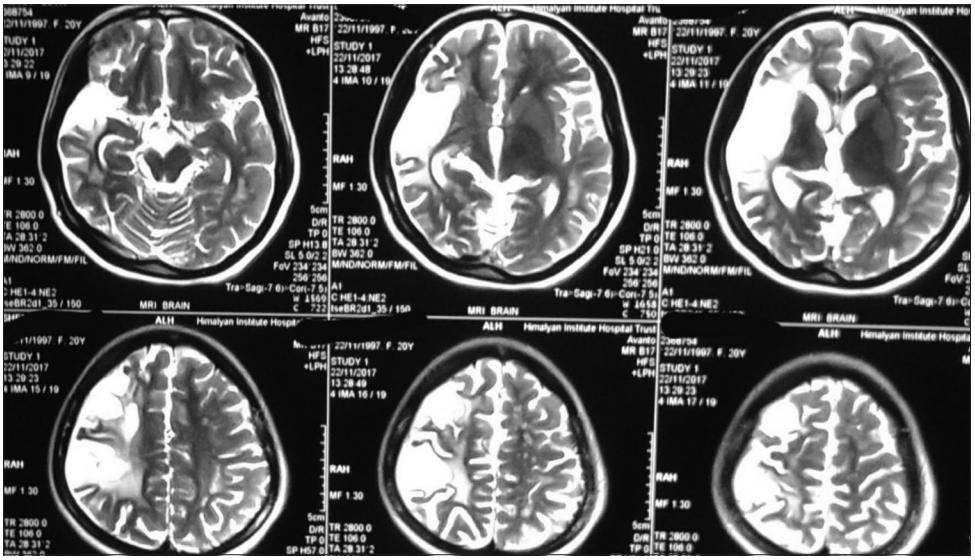

Figure 1: Axial view of magnetic resonance imaging (MRI) brain showing large ill-defined area in the right fronto-temporo-parietal area causing dilatation of the ipsilateral lateral ventricle.

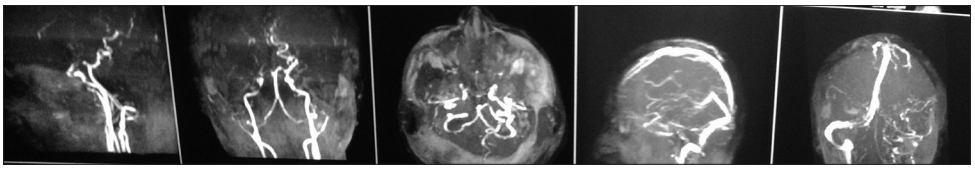

Figure 2: Magnetic resonance (MR) angiogram showing thinned supraclined branches of the internal carotid artery, middle cerebral artery, and anterior cerebral artery with multiple collateral vessels.

branches of the internal carotid artery, middle cerebral artery, and anterior cerebral artery with multiple collateral vessels in the interpendicular and the Sylvain cistern (Figure 2).

Based on history, examination, and laboratory findings, diagnosis of persistent depressive disorder (PDD) with left-sided hemiparesis in partial remission with MMD was made and she was referred to department of neurosurgery for further management.

DISCUSSION

Few important issues in this case are worth discussing: first, she had stroke at young age and psychiatric symptoms started thereafter; second, whether apathy and cognitive decline was related to MMD or congenital rubella infection or levetiracetam; third, whether behavioural symptoms could be explained by depression.

Only few studies have discussed the incidence of stroke in young patients. From the last three decades the reported rates of stroke for individuals aged 15-45 years vary from 3.4 to 21.7 per 100,000,[5] the highest rate observed for Blacks in the Baltimore-Washington area.[6] Common causes of stroke among young is arterial dissection while rare causes include Fabry disease, CADASIL (cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy), MELAS (mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke), MMD, reversible cerebral vasoconstriction syndrome, systemic or primary central nervous system vasculitis, cardiomyopathies, HANAC syndrome (hereditary angiopathy with nephropathy, aneurysms, and muscle cramps), Sneddon's syndrome, sickle-cell disease, and specific coagulopathies.[7]

Our case was suffering from congenital rubella which can rarely progress to PRP or SSPE in which involvement of brain occurs with MRI findings of destruction of white matter, perivascular inflammatory cells and gliosis with moderate neuronal loss, numerous amorphous vascular deposits in the white matter, and severe generalised cerebellar atrophy[8] which are very distinct and differ from that of MMD. Children with congenital rubella infection suffer from neurological and cognitive deficits during early phase of life after which it becomes stable without further determination except in cases of chronic progressive panencephalitis.[9] Our case had delayed milestone and was an average performer in studies till she had an episode of stroke followed by PDD which caused deterioration in her academic performance. There were no features of any panencephalitis in our patient suggesting minimal brain involvement due to rubella.

Many studies have reported cognitive and executive impairment in MMD due to hypoperfusion in the frontal lobes which have been measured with perfusion MRI and the acetazolamide challenge.[10,11] There are evidences available for the development of neuropsychiatric disorders especially in patients with MMD.[12,13] An article in 2012 showed that 37% of the MMD patients presented with significant emotional distress (depression and/or anxiety) while 23% had significant cognitive impairment.[14] Various cortico-striato-thalamo-cortical circuits cross through the striatum and have output in the frontal lobe which might be the reason of widespread motor, cognitive, and emotional effects in MMD.[12]

Patient was receiving levetiracetam for about two years when she reported deterioration in her cognitive functions. Most of the studies indicate that levetiracetam at the right dose and in monotherapy has safe cognitive profile.[15,16] Neurocognitive impairments, such as poor concentration and memory, slowed speed of information processing, and difficulties organising one's thinking (i.e. executive dysfunction), are a central feature of depressive disorder.[17,18] Our patient initially had interest in her studies and was distressed when she was not able to cope at college due to deterioration in her executive and cognitive functioning but later became apathetic showing loss of interest in studies after the development of PDD which caused marked decline in her academic performance.

This case highlights the importance of considering MMD as one of the differential diagnosis while dealing with the young female patient presenting with headache and having a history of hemiparesis whose mother had infection in the antenatal period. It also signifies the presence of psychiatric comorbidities which often are not evaluated in chronic progressive diseases and therefore the need for consultation liaison approach while dealing with such patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the journal. The patient understands that his/her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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