A CASE REPORT OF PUFF OF SMOKE IN THE BRAIN

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Abstract

Moyamoya is a progressive vaso occlusive disease of large intracranial arteries with characteristic collaterals formation. It is more frequent in females compared to males.lt has a bimodal age of onset at around age 5 and at around age 40.The most common initial presentation is Transient ischaemic attack (TIA) is also a frequent initial presentation and maybe recurrent. The symptoms tend to clear rapidly but recurred in some instances. Less commonly,they may present with headache, convulsions, impaired mental clarity, visual disturbance, and nystagmus. In older patients, subarachnoid hemorrhage is the most common initial manifestation.
We report a case of a 38 year old female patient diagnosed with Moyamoya disease.

Keywords: moyamoya disease,vaso occlusive disease,TIA,ischaemic cerebrovascular disease,seizures.

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Case report

We report a case of 38 year old female patient presented with headache followed by tonic clonic movements involving all four limbs,uprolling eyes followed by loss of consciousness and post ictal confusion.Patient had similar complaints of recurrent headache and involuntary movements in the past.On examination vitals were stable.On CNS examination bilateral pupils equally reactive to light, no papilledema .Power ,tone,reflexes and bulk normal and within the normal range ,bilateral plantars flexors and sensory system examination normal. Other systemic examinations were normal.Patient was managed symptomatically and started on Anti epileptics and for headache analgesics were started.Routine investigations were done.[Table 1 and Table 2]

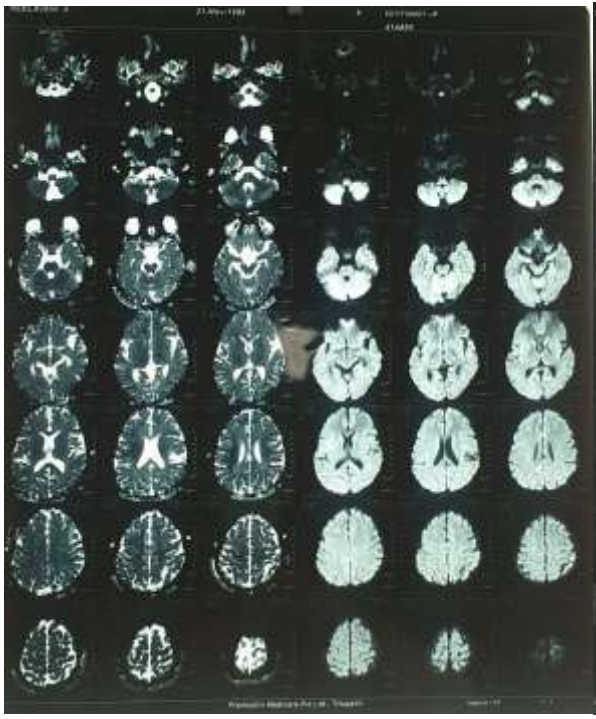
Table 1

Hemoglobin	12.4g/dL
Total count	11,000cells/cumm
Platelet count	1.89 lakhs/cumm
RBS	102 mg/dL
Viral markers	Negative

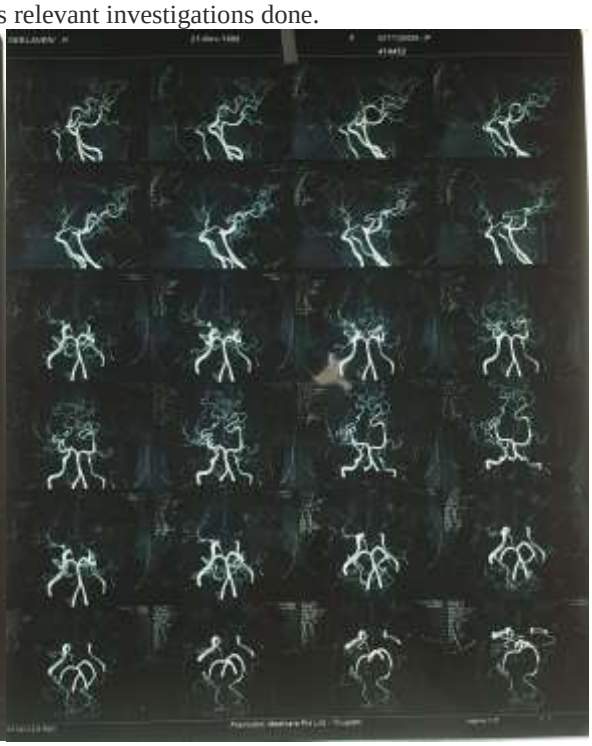
Table 2

PT/INR	13sec/1.1
Serum Sodium	140mEq/L
Serum Potassium	4.0mEq/L
Serum Chloride	109mEq/L
Serum Bicarbonate	22mEq/L

[Table 1] and [Table 2] shows relevant investigations done.



[Image 1]



[Image 2]

MRI brain showed**[Image 1]** small gliotic area in left high parietal lobe suggestive of old ischaemia.MRI Brain Angiogram **[Image 2]**showed moderate diffuse narrowing of cavernous segment of left internal carotid artery with severe narrowing of bilateral MCA and non visualized flow related signal intensity in bilateral middle cerebral arteries with multiple prominent collaterals suggestive of Moyamoya spectrum.(Definitive moyamoya disease)

Our patient has been discharged after improvement with conservative treatment with Anti epileptics and analgesics.As there is no evidence of a potential benefit of antiplatelet use to stroke prevention since the mechanism of MMD does not involve an endothelial damage and thereby platelet adhesion and no other comorbidities anti platelet therapy was not started.Patient recovered well with no neurological deficits in the subsequent visits.

1. Discussion

The Moyamoya disease (MMD) was first described in Japanese literature in 1957. Suzuki and Takaku first named it as "moyamoya disease" in 1969. MMD is an isolated chronic, usually bilateral, vasculopathy of undetermined etiology characterized by progressive narrowing of the terminal intracranial portion of the internal carotid artery (ICA) and circle of Willis.

The overall prognosis is variable. Two-thirds of patients with Moyamoya disease have a symptomatic progression over five years with poor outcomes. Progression of the occlusive process continues regardless of symptom severity, ongoing treatment, age, sex, type and location of the disease.[1]

Moyamoya is a Japanese word for a "haze," "puff of smoke"; it has been used to refer to an extensive basal cerebral rete mirabile—a network of small anastomotic vessels at the base of the brain around and distal to the circle of Willis[2]

Etiology

Inherited conditions and/or association:

- Sickle Cell Disease or trait
- Down Syndrome (Association)
- Neurofibromatosis type 1 (Association)

Acquired conditions:

- Head and/or neck irradiation
- Chronic meningitis
- Skull base tumor
- Atherosclerosis of skull base arteries
- Arteriosclerosis
- Cerebral vasculitis [3][2]

Pathophysiology

The pathophysiology of MMD remains unclear, though genetic predisposition is theorized in East Asian countries. Mutations in BRCC3/MTCP1 and GUCY1A3 genes are implicated in Moyamoya syndrome. Affected individuals are found to have concentric and eccentric fibro cellular thickening of intima within the intracranial portion of ICA. In a study involving the Midwestern US population, an unusually high prevalence of type 1 diabetes, autoimmune thyroid disorders, and other autoimmune disorders were found in the moyamoya cohort which may point towards an autoimmune association. Chronic brain ischemia resulting from the narrowing is believed to be causing an overexpression of proangiogenic factors (fibroblast growth factor and hepatocyte growth factor) which, in turn, would cause the development of a fragile network of collateral vessels [1]

Presentation

The most common initial presentation is Transient ischaemic attack (TIA) is also a frequent initial presentation and maybe recurrent . The symptoms tend to clear rapidly but recurred in some instances. Less commonly,they may present with headache, convulsions, impaired mental clarity, visual disturbance, and nystagmus. In older patients, subarachnoid hemorrhage was the most common initial manifestation.[1]

Characteristics noted in other series have included prolonged TIAs, characteristically induced by hyperventilation or hyperthermia, parenchymal rather than subarachnoid hemorrhages (most situated in the basal ganglia or thalamus), and an unusual "rebuildup" EEG phenomenon in which high-voltage slow waves reappear 5 min after the end of hyperventilation are also seen.[1][2]

Evaluation

1) Magnetic Resonance Imaging (MRI):MRI is one of the first tests to perform in the diagnostic algorithm because it is sensitive and noninvasive. MRI is usually helpful to determine hemorrhages and/or strokes in brain parenchyma. Old ischemic lesions are often seen as white matter hyperintensities in the distal vasculatures and/ border zone areas in FLAIR and T2 weighted sequences. Slowing of flow can be demonstrated by the linear hyperintensities following the sulcal pattern, known as 'ivy sign' in the FLAIR sequence[1]

2) Magnetic Resonance Angiography (MRA):MRA is a gold standard test and provides preliminary information on cerebral arteries and the degree of narrowing. MRA also shows the development of collaterals around the steno-occlusive lesions in the form of 'puff of smoke'. Based on MRA, diagnosis can be formed as either probable or definite moyamoya disease. 'Probable moyamoya' is the presence of a unilateral occlusive process in adults while 'Definite moyamoya' is the bilateral occlusive

process in adults and even a unilateral occlusion in children as the rate of progression from unilateral to bilateral occlusion among children is very high. 'Quasi-moyamoya disease' (also known as Moyamoya syndrome) is a unilateral and/or bilateral occlusive process in the association of any underlying disease. Moyamoya disease does not involve posterior circulation while moyamoya syndrome could involve the steno-occlusive process in the posterior circulation.[1]

2) Conventional cerebral angiography

3) Transcranial Doppler (TCD)

4) Electroencephalography (EEG)

5) Cerebral perfusion measurement[1][4][5]

Apart from the mentioned above, diagnostic tests specific to the other conditions are required if moyamoya syndrome is suspected.[1]

Treatment

The treatment of moyamoya is far from satisfactory. Certain surgical measures have been employed, including transplantation of a vascular muscle flap, omen- tum, or pedicle containing the superficial temporal artery to the pial surface of the frontal lobe temporal pial syn- angiosis with the idea of creating neovascularization of the cortical convexity. These measures have reportedly reduced the number of ischemic attacks, but whether they alter the natural history of the illness cannot be stated. Anticoagulation is considered risky in view of the pos- sibility of cerebral hemorrhage, but there have not been systematic studies.[2]

2. Conclusion

This case highlights the importance of considering Moyamoya disease as a rare differential diagnosis in cases of young females presenting with recurrent headaches and seizures after ruling out other causes for the same. Identifying Moyamoya disease can significantly help in preventing morbidity and mortality and gives an insight into managing the chronic nature of the disease to both the physicians and patients.

Acknowledgement

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