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Moyamoya Disease

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Abstract: *Moyamoya disease is a rare idiopathic progressive vaso-occlusive disorder characterized by irreversible condition of main blood vessels to the brain as they enter into the skull. We present a case of 48 years old Indian female presenting to the Hospital with sudden onset of loss consciousness followed by facial deviation to the left side with weakness on right half of the body, also associated with headache. The patient was investigated with CT Angiogram, diagnosed as moyamoya disease.*

I. Background

Moyamoya disease is a rare progressive vaso-occlusive disorder of an unknown etiology. It is characterized by progressive stenosis of terminal portion of internal carotid arteries bilaterally and the main track of Anterior and middle cerebral artery and is associated with collateral vessels at the base of the brain. When similar clinical manifestation are associated with an underlying disorder, it is referred to as moyamoya syndrome. Since the diagnostic criteria of the disease are mainly based on angiographic findings, it is recommended that the term moyamoya syndrome should avoided. The pattern of presentation of moyamoya disease report in the literature so far have shown several consistent features, such as hemiparesis, monoparesis or sensory disturbances reflecting TIA or intra cerebral intra ventricular and subarachnoid haemorrhage in adults whether the particular pattern of presentation are universal.

II. Case description:

A 48 year old Indian woman presented to the hospital with sudden onset loss of consciousness while cooking at home which lasted for 1 hour. On regaining consciousness patient had facial deviation to left side with weakness on the right half of the body. Patient had complaints of headache since 5 to 6 years which was of moderate intensity and piercing type in nature and was associated with nausea and vomiting, it used to persist for few hours and not relieved by NSAIDs. She had no history of concurrent fever, trauma, convulsion, involuntary movement, change in speech, sensory impairment, dysphagia, neck stiffness or vision problems. On evaluation, the patient stated that she had similar episode 11 years back when she was diagnosed as a case hemorrhagic stroke which was probably caused by hypertension. She had stopped her anti-hypertensive medication after 1 month and since then she is irregular on treatment. She had history of pulmonary tuberculosis in 1973 for which she was on treatment for 4-5 years. Her family history was significant for hypertension to both parents. The patient denied any family history of stroke, seizures or cancer. Socially, she was a housewife. She didn't consume alcohol, use illicit drugs or smoked cigarettes.

On physical examination, the patient was conscious, co-operative and oriented to time, place and person. She was thin built with BMI of 20. Vital signs (Blood Pressure, 154/92 mm Hg; Heart rate 62beats per minute; Temperature 98.6 ^{0}F, Respiratory Rate 18/minute) were normal. Neurological examination showed motor weakness on right half of body with strength 3/5 in upper and lower limb with extensor right plantar, diminished reflexes and normal muscular tone. All cranial nerve except 7th on right side were intact. Respiratory examination revealed inspiratory crepts over the left side upper lobe. Cardiovascular and per abdominal examination were unremarkable.

Complete blood count showed normocytic anemia with normal leukocyte and platelet counts. Coagulation profile was within normal limits and hypercoagulable workup was not significant. Specific test for hypercoagulability disorders included activated protein C, antithrombin III, homocystiene, D dimer, Factor V Leiden, fibrinogen, lupus anticoagulant, partial thromboplastin time, protein S, Prothrombin time and thrombin time. Results were normal for all these studies. ANA (1.9) and Anti-Cardiolipin Antibodies IgG (65.3) were positive.

A CT angiogram of brain revealed hyper dense intra-parenchymal hemorrhage with perifocal edema seen in left basal ganglia and the hemorrhage is transecting into bilateral lateral ventricles, 3rd ventricle and 4th ventricle. Significant luminal stenosis seen in supraclinoid part of Internal Carotid artery, proximal part of bilateral Middle Cerebral Artery and bilateral Anterior Cerebral Artery with multiple collateral formations predominantly in bilateral basal ganglia region consistent with MOYAMOYA Disease. Despite the

cerebrovascular disease manifested radiologically, the patient did not exhibit any decline in cognitive function. Patient was managed symptomatically with anti hypertensive and NSAID's. The patient continues to follow up on OPD basis and is stable and stroke free.

III. Discussion

Moyamoya disease is a noninflammatory and nonatherosclerotic condition characterized by chronic, progressive occlusion of the Circle of Willis artery that least to the development of the characteristic collateral vessels seen on imaging particularly cerebral angiography. Moyamoya was originally considered to affect predominantly persons of Asian heritage but has now been observed throughout the world.Moyamoya usually present recurrent headaches which is migraine like in quality and refractory to medical therapy. 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 carotid which looks like puff of sense. Though MRI angle is used to confirm the diagnosis and to seen the anatomy of the vessels involved, CT-Angiography Brain can also be used to see intracranial stenosis suggesting moyamoya.

Differential diagnosis:

Differential diagnosis of headache with hemiplegia includes arterio-vascular malformation with steal phenomenon and Transient Ischemic Attack.

Treatment:

Various medical management of moyamoya disease gave been researched. Anticoagulant and antiplatelet agents have shown no remarkable benefit. The lack of obvious efficacy has been described for corticosteroid in moyamoya disease.Patients are also treated symptomatically with non-steroidal anti-inflammatory drugs, propranolol and prochlorperazene with variable response. Surgical treatment modalities have been used to manage the hemorrhage and ischemic consequences of moyamoya disease. Ventricular drainage and hematoma evacuation are often useful for hemorrhage cases. In ischemic moyamoya disease surgical methods have been used to restore and maintain adequate cerebral perfusion.Serious adverse effect of Surgical intervention in moyamoya disease includes death, cerebro-vascular infarct or hemorrhage, motor or sensory dysfunction and cognitive decline.

Prognosis:

Moyamoya disease has a more rapid and worse progress in children younger than 3 years than in than in those aged 3 years or older. However when comparing children with adults the prognosis in generally worse for adults because that have increased hemorrhagic episode and thus a higher mortality.

IV. Conclusion

A better understanding of the natural history of patient with moyamoya diseases as well as the benefit of the various treatment modalities is needed. Ideally structural randomized clinical trials would held to provide insight into the optimal treatment methods for the patient. Management of moyamoya disease remains ill-defined and varies depending on the inherit provisions of individual institution. Careful long term neurologic and radiologic follow up is essential in adult patient with moyamoya disease to prevent additional stroke events and improve outcome.

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