ejpmr, 2016,3(11), 573-575

SJIF Impact Factor 3.628

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

"A SICKLE CELL TRAIT CHILD WITH A SEQUELA -MOYAMOYA SYNDROME"

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Article Received on 13/09/2016 **Article Revised on 03/10/2016** **Article Accepted on 24/10/2016**

ABSTRACT

Moyamoya Syndrome is a chronic cerebrovascular disorder characterized by progressive steno-occlusion of cerebrovascular arteries leading to development of collaterals network at the base of brain. It has been seen as an uncommon neurological complication of few haemoglobinopathies including sickle cell disease, however only three such cases of **moyamoya syndrome with sickle cell trait** have been reported in the literature. We report a 6 yr. old female child, who presented to hospital with headache and vomiting, subsequently developed sudden loss of vision and symptoms of neurological dysfunction diagnosed as moyamoya syndrome associated with her known **sickle cell trait etiology.**

KEYWORDS: headache, sickle cell trait, moyamoya syndrome.

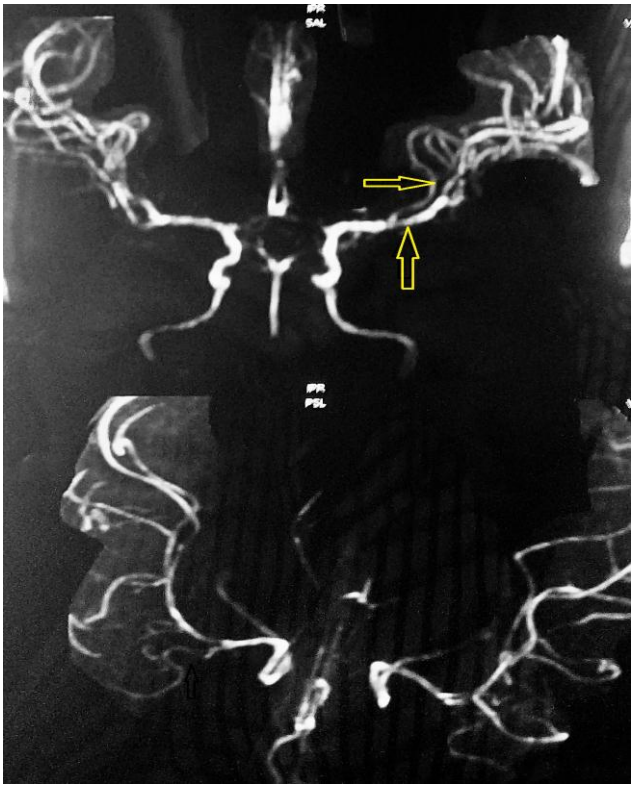
INTRODUCTION

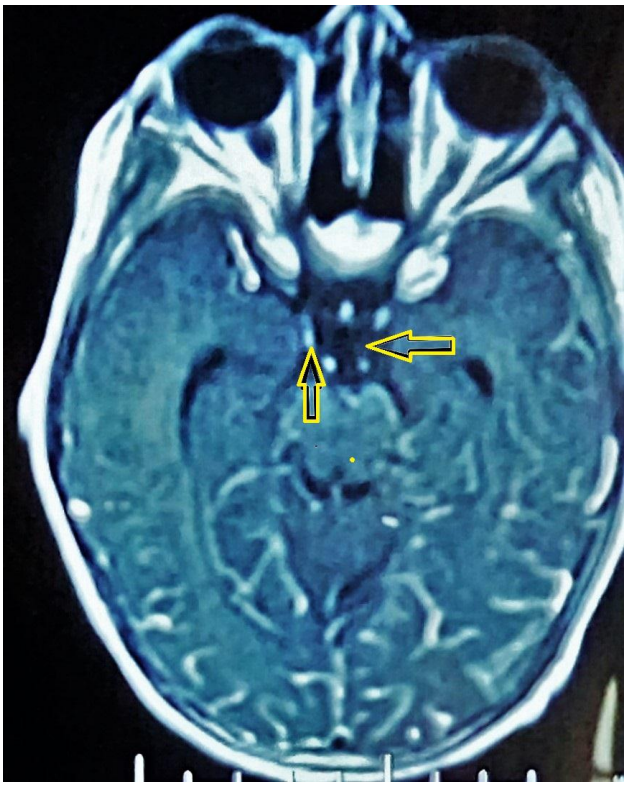
Moyamoya Syndrome is a chronic cerebrovascular disorder characterized by progressive steno-occlusion of cerebrovascular arteries leading to development of abnormal vascular network (collaterals) at the base of brain[1,2]. The etiopathogenesis is unknown, however environmental, genetic and infections (viral, bacterial) are the major risk factors.[1,3] The disease manifests as recurrent transient ischemic attacks, seizures, headache, visual defects and stroke in children. **Moyamoya syndrome** has been seen as an uncommon complication of few haemoglobinopathies including sickle cell disease, however only three such cases of moyamoya syndrome with **sickle cell trait** have been reported in the literature.[4,5,6] In this paper, we reported a known case of sickle cell trait with a sequelae moyamoya syndrome as its uncommon neurological sequelae.

CASE REPORT

A 6 yr. old female child of rural origin from central India was brought to our Pediatrics department with chief complaints of headache and vomiting since 3 days, as per the parents headache was severe and associated with night wakefulness. The child was also admitted 13 days back with complaints of pain in lower limbs and chest and investigated thoroughly with complete blood count and haemoglobin electrophoresis, reports were suggestive of sickle cell trait as AS pattern .The child was treated and discharged with necessary advice. On general physical examination, child was irritable, oriented, pupils were round reactive equal in size, normal tone/ power in all four limbs, deep tendon reflexes and plantar reflexes were normal. There were no signs of meningeal irritation and cerebellar dysfunction. The child was investigated and laboratory reports on admission were haemoglobin 8.6 mg/dl, white blood cell 15,900/mm3, platelet 1,84,000/mm3. On the next day child developed sudden loss of vision associated with headache, fundus examination was suggestive of papilledema with features of raised intracranial tension on examination with mid dilated unequal pupils and positive McEwan sign ; MRI with MR Angiography was done in our hospital and it revealed findings suggestive of moyamoya syndrome with markedly attenuated bilateral supraclinoid internal carotid and middle cerebral artery with multiple small collaterals and acute infarct in grey ,white matter of high anterior frontal lobe, bilateral posterior tempro-parital lobe and bilateral occipital lobe. Underlying etiology was investigated with necessary sampling as renal function test, connective tissue disorder[ANA], liver function test was done and found normal.

The patient is a diagnosed case of sickle cell trait [AS pattern], now presented with a neurological sequela as moyamoya syndrome, after prompt management and stabilization child was referred to higher neurological centre for further management as per parents request.





DISCUSSION

Moyamoya is radiologically characterized by chronic progressive intracranial arterial occlusion with narrowing of distal internal carotid arteries and anterior and middle cerebral trunks leading to development of fine network of collateral channels at the base of brain.[7] However, the etiopathogenesis is unclear but the factors like environmental, genetic and infections play an important role in the origin.[1,3] Moyamoya syndrome [puff of smoke or haze] is tangle of tiny vessels which has been noted in variety of ethnic groups and is found as a nonspecific response to various underlying conditions like sickle cell anaemia, protein C, S deficiency, head trauma, tuberous sclerosis, Marfan syndrome, previous bacterial or tubercular meningitis.[7] Thus, the term moyamoya disease should be used in cases with characteristic MR angiographic findings with unclear etiology and moyamoya syndrome in the children with a known etiology.

Sickle cell disease is a recognised cause of thrombotic cerebral infarcts and moyamoya is an uncommon neurological complication of sickle cell disease. A study of 44 patients maintaining chronic transfusions observed that risk of recurrent stroke was higher in patient of moyamoya than those without disease.[8] Inspite of significant association between moyamoya and sickle cell disease(HbSS), the relationship is rare in benign conditions like **sickle cell trait**[9], only three such cases have reported in literature till now.[4,5,6] A case report of Mustafa et al reported 7 year old boy presented with complaint of headache and diagnosed as moyamoya syndrome associated with sickle cell trait[6], similarly Agrawal et al reported a cases of 44 year old man with complains of numbness and weakness of left side of body, MRA revealed bilateral moyamoya syndrome with sickle cell trait as a underlying etiology.[5] In some case reports sickle cell trait has been found associated with cerebrovascular complications including stroke. A study of 21 children with sickle cell trait with brain abnormalities was done and it was found that children with arterial torturing have significantly greater ratio of HbS than that in controls.[10] Sickle cell trait patients have membrane defect of sickle cell erythrocytes due to loss of normal phosphate lipid symmetry, resulting abnormal phosphatidyl serine exposure leading to haemostatic perfusion and finally increasing venous thromboembolic events.[11]

The symptomatology of moyamoya syndrome has varied presentation as mentioned above with headache as first and commonest symptom[1], however recurrent ischemic attacks and ischemic strokes can occur in paediatric moyamoya syndrome. A study of Scott et al[12] reported 67.8% stroke and 6.3% headache, similarly in Guzman et al[13] study observed majority of cases with ischemic attack (51%) followed by headache (44%).[13]

Various previous reports with worse outcome and sudden death with sickle cell trait, leads to a vision that moyamoya syndrome is an uncommon complications of sickle cell trait and should be kept in mind with cases of neurological disturbances and MRI should be performed in cases of sickle anaemia coming from high risk prevalent areas.

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