ORIGINAL RESEARCH PAPER

Paediatrics

MOYAMOYA DISEASE IN A CHILD : A RARE CASE

KEY WORDS:

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ABSTRACT

- One of the rare causes of ischemic stroke being Moyamoya disease which is very rare in children.
- Here we report a case of 6 year old female child admitted to hospital with transient ischemic attacks and right sided hemiparesis.
- On MR Angiography, child was diagnosed to have Moyamoya disease.This child was treated in our institute conservatively and was referred to higher centre for specific neurosurgery.

INTRODUCTION:

- Moyamoya disease which was first reported in Japan in 1957 and it's incidence is higher in Asian people[1].
- It is an occlusive cerebro vascular disorder caused by blockade of internal carotid artery at the base of brain around basal ganglia.
- There will be oxygen and blood deprivation to brain due to blockage of vessels which leads to signs and symptoms like seizures, headache, loss of speech, visual disturbances, stroke ,transient ischemic attacks.
- The classical pediatric presentation is recurrent transient ischemic attacks or complete ischemic strokes.
- Surgical revascularization remains the main stay of treatment to improve long term outcome .
- If not treated , can result in permanent neurological and cognitive deficits.
- Because of the rarity of the disease and of clinical interest, we are reporting the case of Moyamoya disease.

CASE REPORT:

- A 6 year old female child born of non consanguinous parents presented with history of suddenly developed weakness of right side of the body with aphasia since 8days.
- No history of fever, seizures, head injury, ear discharge.
- Child has past history of left side hemiparesis 8 months prior to admission. Developmental history of child is normal.
- On examination, child is conscious , coherent , afebrile and mildly anemic. No neurocutaneous markers or asymmetry of face.
- On neurological examination, hemiplegic gait, decreased tone in right upper and lower limb, power was 2/5 over right upper and lower limbs, deep tendon reflexes are exaggerated, plantars were extensors on right side.

INVESTIGATIONS:

1)Hematological- Hemoglobin: 8g/dl. No thrombocytosis .
2)Prothrombotic profile:

- Factor V Leiden mutation: not detected
- Protein C activity: 104% (normal)
- Protein S activity: 13% (low)
- Antithrombin activity: 127% (high)
- Lupus anticoagulant: absent
- Cardiolipin IgG - 36.6 GPL/ml(high)
- Cardiolipin IgM- 1.90 MPL/ml (normal)
- Anti B2 glycoprotein 1 (IgG and IgM)- normal.

3) coagulation profile:

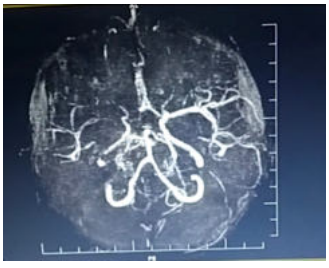
- PT: 15seconds
- APTT: 32seconds
- INR: 1.2

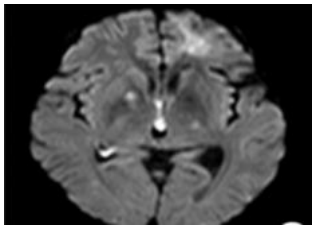
4) Renal function tests, liver function tests, serum electrolytes were normal.

5) 2D ECHO and ECG was normal.

6) MRI BRAIN: Left frontoparietal lobe acute infarct with right frontoparietal lobe chronic infarct with gliotic changes and volume loss in right frontoparietal region.

7)MR ANGIOGRAPHY: Diffuse long segment reduced caliber noted in right ICA and right MCA causing about 80-90% stenosis suggestive of Moyamoya disease.





TREATMENT GIVEN

- We treated child conservatively with oral Aspirin @5mg/kg/day. Child showed slight improvement in power on right side(3/5).
- As our institute has no equipment for neurosurgery, we referred the child to higher centre where cerebral revascularization surgery using Encephalo Dural Arterio Synangiosis(EDAS) was done.

FOLLOW UP

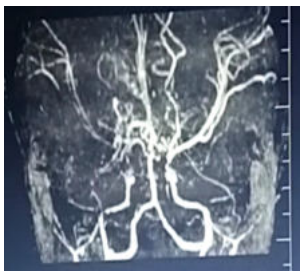
- Child came for follow up and has shown improvement after surgery and repeat MRI was found to be normal.

DISCUSSION:

- Mean age at which Moyamoya disease diagnosed is

6years with range between 6months to 8years.

- In this disease, MRI not only reveals the infarction, but also visualises the collateral vessels as multiple blood flow voids at the base of brain and basal ganglia.[2]
- MR angiography shows classical "Puff of smoke appearance".[3]



- Children who are managed conservatively experienced stroke at a rate ranging from 3.2-15.0%, but after surgical intervention it is usually 0.0-1.6%.[4]
- So, surgical management is preferred.

CONCLUSION

- Definitive management for Moyamoya disease has not determined yet. As it is a progressive disease , surgical management should be considered in symptomatic patients.
- Early diagnosis and management is mandatory to prevent further hemorrhage and stroke.

CONFLICTS OF INTEREST: No

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