

Acetazolamide Challenge ^{99m}Tc ECD Brain Perfusion SPECT in Moya Moya Disease

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Introduction

- Moya Moya disease (MMD) is a chronic, idiopathic, bilateral, progressive steno-occlusive disease typically involving the intracranial ICAs and cerebral arteries, results in ischemic or haemorrhagic events
- Typical appearance on cerebral angiography i.e; "puff of smoke" and refers to the appearance of multiple compensatory dilated striate vessels seen on angiography
- CT and MRI play a major role in documenting the regions of infarction/hemorrhage
- This study was performed to analyze the role of brain perfusion SPECT and Acetazolamide challenge in diagnosis and management of Moya Moya Disease

Materials and Methods

- Retrospective analysis of the records of patients with Moya Moya disease referred for Acetazolamide brain perfusion SPECT between Feb 2021 to June 2022, was performed
- Ten patients of age range 5-65 yrs were included in the analysis, out of which 6 were male and 4 were female patients
- Five children of age group 5 to 16 years and 5 adults between 30 to 65 years
- All patients underwent baseline & post acetazolamide ^{99m}Tc-ECD brain perfusion SPECT on two separate days
- Diagnosis of Moya Moya disease was established on basis of typical vascular pattern on DSA in 2 patients, on CT angiography in 1 patient, MR angiography in 7 patients
- All patients underwent baseline & post acetazolamide ^{99m}Tc ECD brain perfusion SPECT as per the established procedure guideline to assess cerebro-vascular reserve
- One patient had a follow up brain perfusion scintigraphy at 3 months after the first scan. Two patient underwent surgical procedure (EDAMS)

Image Acquisition

- Image acquisition was done at 40 min after injection of 20 mCi ^{99m}Tc ECD under dual headed gamma camera coupled with HRGP collimator. Images were acquired in 128 x 128 matrix over 360 degree continuous, rotation acquiring 64 frames, each frame of 20 sec duration
- Acquired data were processed and attenuation correction method was applied & images were displayed in transverse, sagittal and coronal sections
- Visual interpretation of the perfusion state was made using a rating scale of 1 to 5, in which; 1 mild 2 moderate & 3 severe reductions in perfusion, 4 normal & 5 increased perfusion

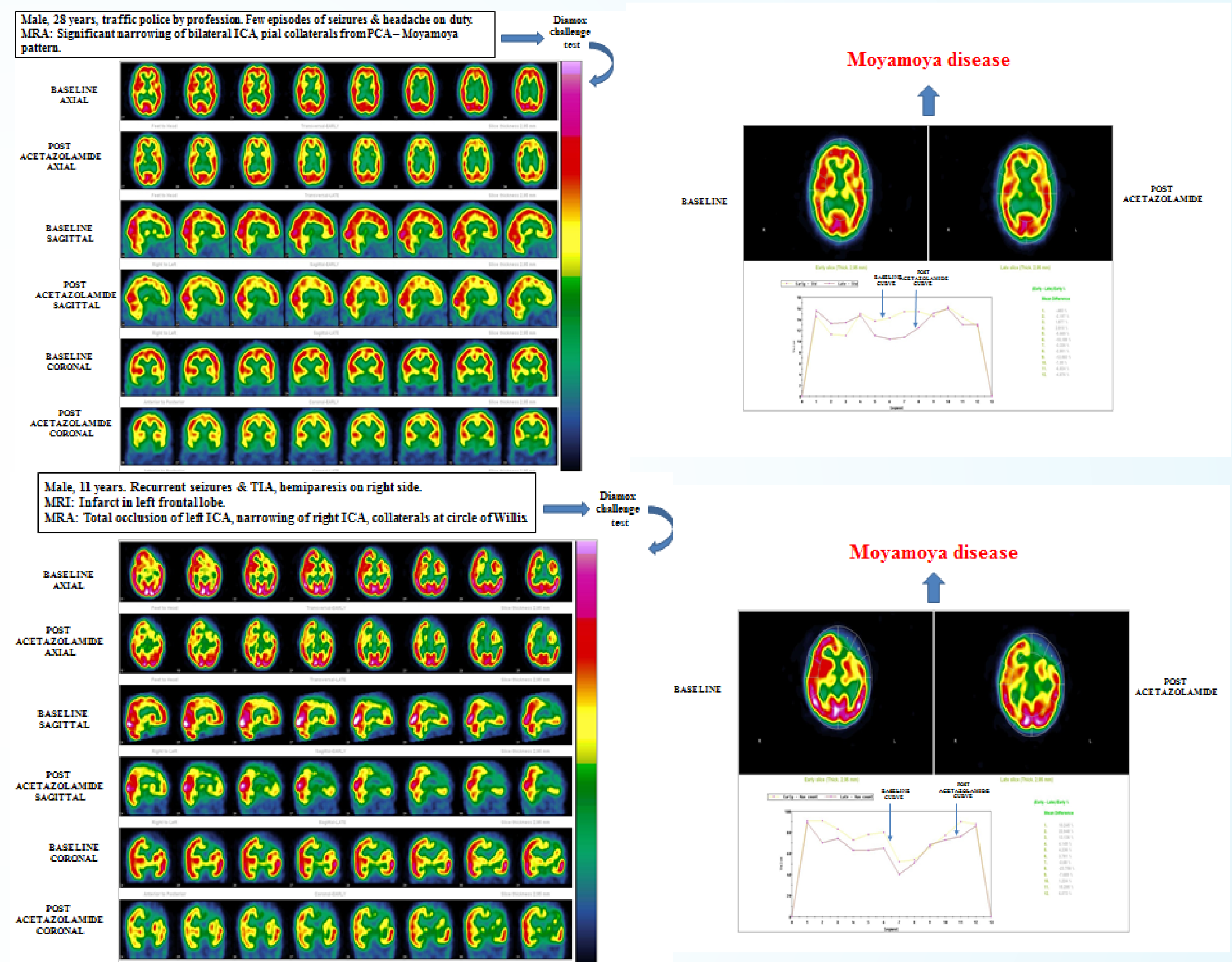
Results

- Patients were referred for brain perfusion scintigraphy at a varying duration of 3 days to 2 months after the symptom onset
- Six patients presented with hemi paresis, two with seizures, four with headache, five with transient ischemic attack and one with stroke
- Diagnosis of Moya Moya disease was established on basis of typical vascular pattern on DSA in 2 patients, on CT angiography in 1 patient, MR angiography in 7 patients
- Three patients had evidence of parenchymal infarcts on MRI
- Five patients underwent vessel wall MRI, of which three were normal & two showed abnormalities
- Visual scoring of brain perfusion scintigraphy images was performed in baseline and post acetazolamide studies
- For analysis brain was divided in 12 areas 6 on each side
- Total 120 areas of brain were analysed. Normal/preserved perfusion is seen in 78/120 areas in baseline study and 72/120 in post ACZ study. Mild, moderate & severe defect in perfusion is seen in 10, 16 & 16 areas in baseline whereas 16, 6, 26 areas in post acetazolamide study. 44 areas in post acetazolamide study showed increase in perfusion compared to baseline
- Change in perfusion in post ACZ SPECT was categorised to three types of response according to Rogg's criteria . 63, 13 & 39 areas were categorised in type I,II & III categories respectively

TABLE SHOWS NUMBER OF AREAS SHOWING NORMAL AND ABNORMAL PERFUSION AT THE BASELINE AND POST ACETAZOLAMIDE STUDY ALONG WITH TYPE OF RESPONSE POST ACETAZOLAMIDE

BRAIN REGIONS	BASELINE		POST ACETAZOLAMIDE						RESPONSE (ROGG'S CRITERIA)						
	NORMAL	ABNORMAL (REDUCTION)	ABNORMAL (REDUCTION)			PRESERVED			ABNORMAL (REDUCTION)						
			MILD	MODERATE	SEVERE	NORMAL	INCREASED	ABNORMAL	I	II	III				
RF	3	7	3	3	1	3	1	2	7	1	2	4	3	1	6
RP	6	4	1	2	1	4	2	2	6	2	0	4	3	1	6
RT	9	1	0	1	0	7	2	5	3	2	0	1	7	0	3
RO	10	0	0	0	0	10	5	5	0	0	0	0	10	0	0
RBG	10	0	0	0	0	8	3	5	2	2	0	0	8	0	2
RTHAL	10	0	0	0	0	8	2	6	2	2	2	0	8	0	2
TOTAL RIGHT	48	12	4	6	2	40	15	25	20	9	2	9	39	2	19
LF	1	9	3	2	4	2	1	1	8	0	2	6	1	1	8
LP	2	8	2	2	4	2	2	0	8	2	2	4	1	4	5
LT	3	7	1	4	2	4	2	2	6	3	0	3	3	4	3
LO	3	7	0	0	3	7	4	3	3	1	0	2	7	1	2
LBG	8	2	0	1	1	8	2	6	2	1	0	1	8	1	1
LTHAL	9	1	0	1	0	9	2	7	1	0	0	1	9	0	1
TOTAL LEFT	26	34	6	10	14	32	13	19	28	7	4	17	29	11	20
OVERALL TOTAL	74	46	10	16	16	72	28	44	48	16	6	26	68	13	39

RF-RIGHT FRONTAL, RP-RIGHT PARIETAL, RT-RIGHT TEMPORAL, RO-RIGHT OCCIPITAL, RBG- RIGHT BASAL GANGLIA, RTHAL-RIGHT THALAMUS; LF- LEFT FRONTAL, LP- LEFT PARIETAL, LT- LEFT TEMPORAL, LO- LEFT OCCIPITAL, LBG- LEFT BASAL GANGLIA, LTHAL- LEFT THALAMUS (NUMBER DENOTES INVOLVED AREAS OF BRAIN OUT OF TOTAL PATIENTS)



Discussion

- Attention should be given to Rogg's type III and Kuroda's type 2, as this is the most important one to find out and we would have missed without acetazolamide challenge
- In clinical routine, this type of normal basal perfusion and reduced vascular reserve are so prevalent
- Once basal perfusion starts to reduce, there is the chances to progress further or if left without any treatment, patients would go the natural disease course while suffering from a new stroke episode
- On basal/acetazolamide SPECT, cerebral ischemia will be evaluated as preserved or reduced vascular reserve to acetazolamide challenge
- Once basal perfusion begins to reduce (Kuroda's type 3), CT perfusion or MR perfusion studies or arterial spin labeling MR might be able to find the basal hemodynamic failure

Conclusion

Acetazolamide brain perfusion SPECT has a useful role in assessment of cerebro-vascular reserve in management of patients with Moya Moya disease & helps in selecting suitable patient for cerebrovascular anastomotic surgery