

A CASE OF RECURRENT ALTERNATING TRANSIENT HEMIPARESIS

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DNB Resident

Railway Hospital, Perambur.

6 year old school going child.

Apparently normal till 3 yrs when she developed

In EVENING:

Rt. focal clonic seizures

- 2 episodes
- In 1 hr interval
- Each lasting 5 min
- Angle of mouth to Rt.
- No loss of consciousness
- No limb weakness

➤ Advised to go to higher centre by a Pvt. practitioner.

Next day MORNING: Woke with

Rt. sided limb weakness.

Loss of speech

Admitted in Pvt hospital in Chengalpet

CT brain – Infarct in Lt. FrontoParietal region.

ECHO Heart

Coagulation Profile

Lipid Profile

Baseline Inv (CBC,LFT,RFT) - NORMAL

NORMAL

E
0mm
OD
1.4
02, 248)

24.01.08 13:47:17.
120kV/240mA



In a week

- Speech improved in 6 days with slight stuttering.
- Minimal distal weakness continued.

In ICH, Egmore:

MRI BRAIN - Similar findings.

Aspirin, Carbamazepine – advised.

MR A30
HFS
+LPH
09-Feb-05, F, 3Y
STUDY 1
09-Feb-08
2:04:30 PM
5 IMA 11 / 15

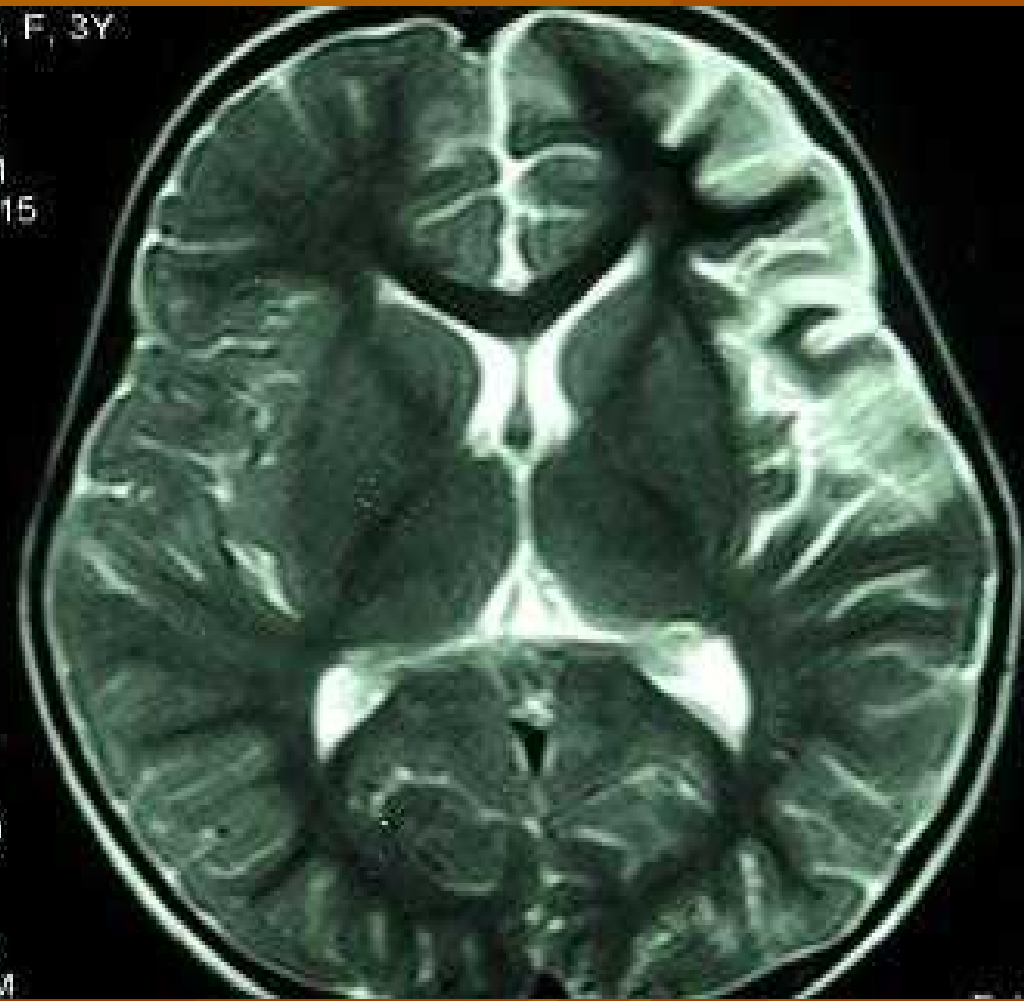
MR A30
HFS
+LPH

5cm
RHA

MF 1.29
E TR 5550.0
TP 0 TE 104.0
H48.6 TA 39.12
SL 5.0 BW 200.0
941230 M/ND/FSM

5cm

E
TP 0
SP H55.1
SL 5.0



New symptoms

2 ½ years later

5 months ago...

Left sided weakness – sudden onset; in MORNING

Not able to walk.

Unable to hold objects in Lt hand.

Lasted for 5 min → then improved.

No loss of consciousness.

Similar episodic weakness – in EVENING

Same side

Now speech also affected.

Lasted 10 min --> then improved.

Admitted in our Hospital.

O/E

- Conscious
- Oriented to time, place & person
- Comfortable

Vitals: Normal.

No Neurocutaneous markers seen.

CNS Examination

Higher Mental Functions:

Speech – Aphasic

Cranial Nerves:

7th(Lt) – UMN type Facial palsy

Motor System:



- Bulk
- Tone

Normal

➤ Power

- UL - 3/5 (L)
- 4/5 (R)
- LL - 5/5 (R & L)

Reflexes:

- Superficial : Present; Plantar – Left 
- Right - 
- DTR : Exaggerated (L) ; Normal (R)

No involuntary movements.

Gait – Hemiplegic gait.

Spine and Cranium – Normal.

Bladder and Bowel – Normal.

CVS, RS, Abdomen - Normal.

Provisional Diagnosis :
Rec. Alternating Transient
Hemiparesis

DD

1. Procoagulant states.
2. Carotid thromboembolic disease
3. Blood dyscrasias and stroke
4. Vasculitides
5. Basilar artery thrombosis
6. Metabolic disorders (MELAS, Homocysteinuria, Methyl malonic acidemia, Fabry disease)
7. Cerebral Aneurysms

INVESTIGATIONS

- Pr C, Pr S, AT-III, Factor V Leiden – Normal.
- AntiPhospholipid, Anticardiolipin, Lupus anticoagulant antibodies – Negative.
- Blood Lactate – Normal.
- Carotid Doppler – Normal.
- PH – Normal
- ESR – Normal
- Serum homocysteine levels - Normal

CT angiography - Revealed the Diagnosis

Aarthi Scans Parameswari. 6/F
m Railway., 1289
inition AS+ *28-Jun-2005, F, 6Y
CT 2010B 28-Jun-2011
15:07:47.11
608 IMA 1
Manip, MIP

HAL

128 Slice CT, Aarthi Scans
Ref.: Southern Railway
Definition AS+
CT 2010B

Spin: 2
Tilt: -23

RHA
5cm



MCA

ICA

BA

ICA

ICA

ECA

ECA



5cm

- The petrous, cavernous and supraclinoid parts of **right ICA** appears thin calibered.
- **Left ICA** reveals critical stenosis beyond its P1 segment. Distally reformed by collaterals.
- Anterior cerebral arteries appear normal on both sides.
- Basilar artery and both vertebral arteries are normal.
- *'Puff of Smoke'* appearance of Collaterals.

28-Jun-2011
15:07:47.11
606 IMA 1
Manip, MIP

Definition AS+ *28-Jun-2005, F, 6
CT 2010B 28-Jun-2011
15:07:47.11
606 IMA 1
Manip, MIP

Spin: 2
Tilt: -23

RHA

RHA

5cm

Parameswari, 6/F
1289

HLA

128 Slice CT, Aarthi Scans Parameswari, 6/F
Ref.: Southern Railway., 1289

HLA

128 Slice CT, Aarthi Scans Parameswari, 6/F
Ref.: Southern Railway., 1289
Definition AS+ *28-Jun-2005, F, 6Y

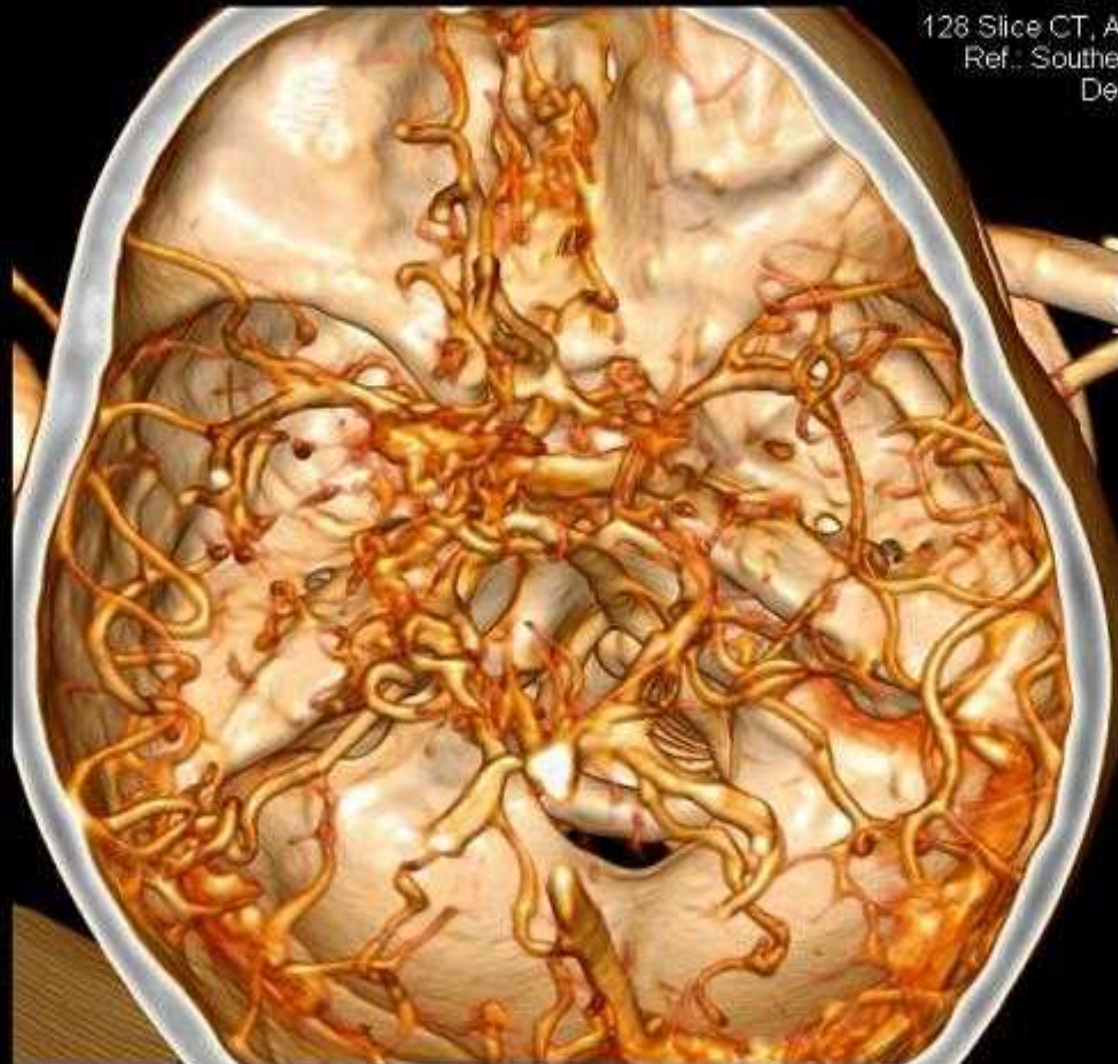


W 682
C 343

Parameswari. 6/F
ID: 1289
* 6/28/2005
Study 1
6/28/2011
3:07:43 PM
1 IMA

128 Slice CT, Aarthi Scans
Ref.: Southern Railway,
Definition AS+

Spin: -177
Tilt: 113



Comment: CTA.

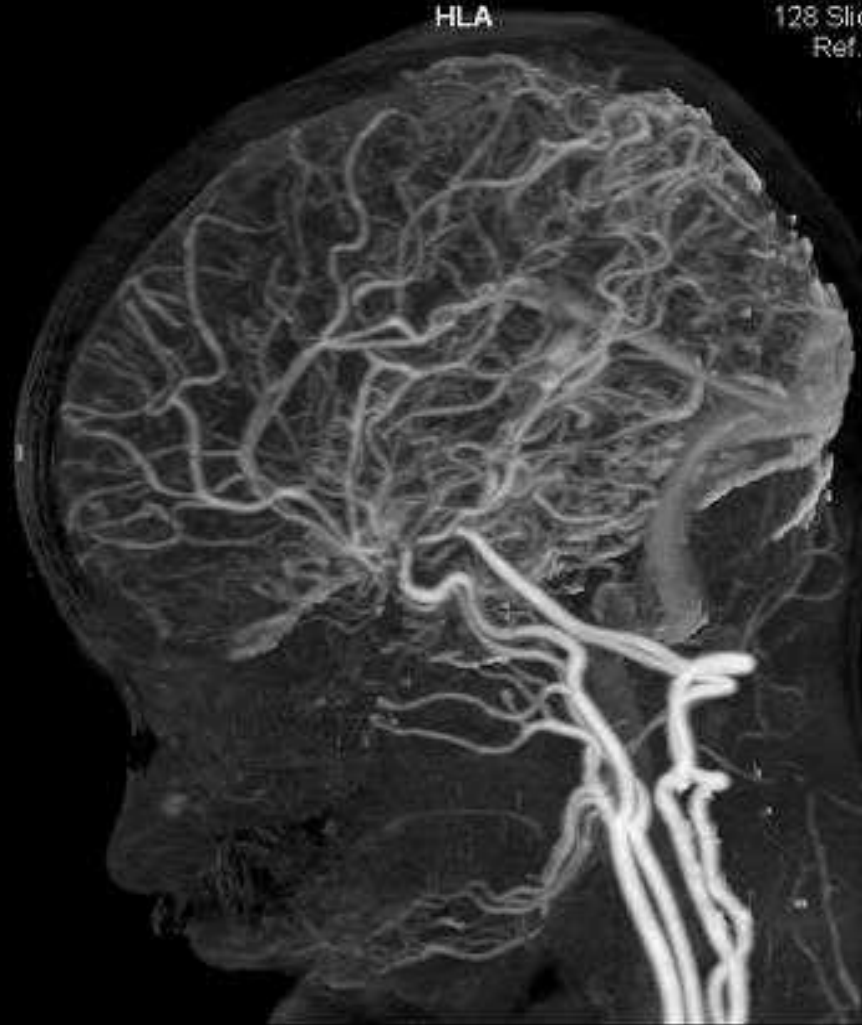
Parameswari, 6/F
ID: 1289
* 6/28/2005
Study 1
6/28/2011
3:07:47 PM
1 IMA

HLA

128 Slice CT, Aarthi Scans
Ref.: Southern Railway,
Definition AS+

Spin: -93
Tilt: -1

ALF



C APPLIED
Comment: slim bone mask



W: 682
C: 343

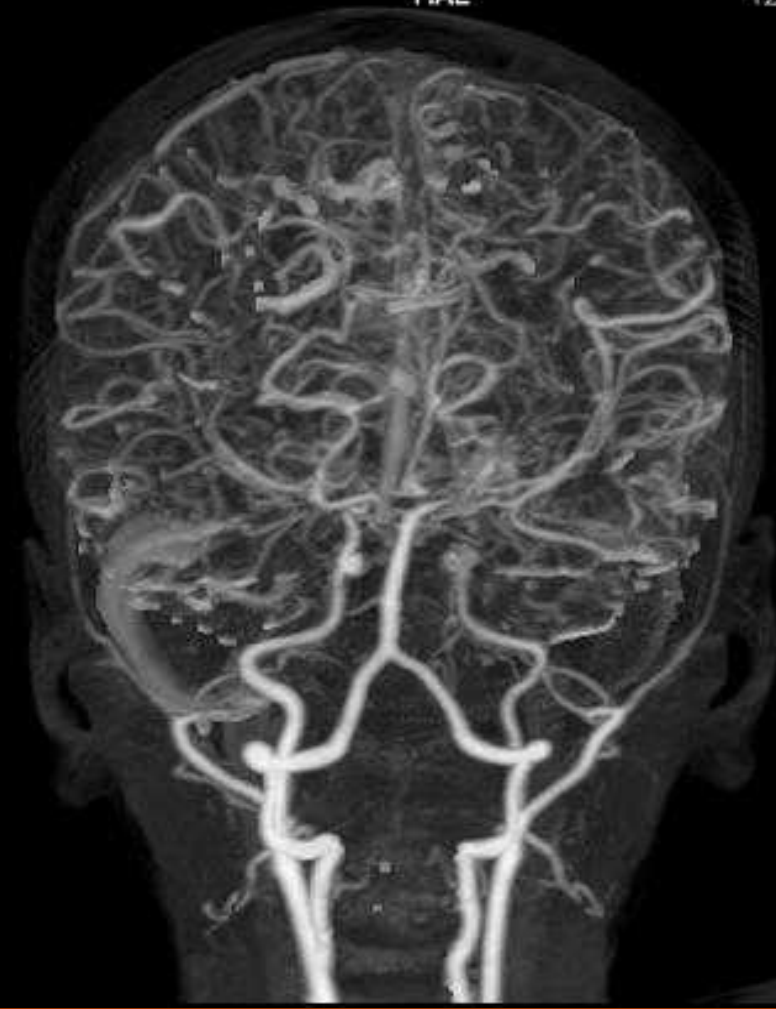
Parameswari, 6/F
ID: 1289
* 6/28/2005
Study 1
6/28/2011
3:07:47 PM
1 IMA

HAL

128 Slice CT, Aarhi Scans
Ref.: Southern Railway,
Definition AS+

Spin: 2
Tilt: -23

RHA



C APPLIED
Comment: slim bone mask

W: 682
C: 343

Diagnosis

MOYA-MOYA DISEASE

Neurosurgical intervention: (VHS,Adyar)

- **Revascularization procedure**
(*Galeo arachnoid synanastamosis*) with multiple burr holes done.
- Carbamazepine and Aspirin continued.
- Physiotherapy and Speech therapy continued.

Presently...

Able to walk.

Able to eat with Rt. hand with minimal spilling.

Left side weakness improved.

Continues to be Aphasic.

Moya-Moya Disease

Moya moya disease was first described in Japan in 1957.

Progressive steno-occlusive disease at terminal portions of the bilateral internal carotid arteries with the development of "moyamoya vessels" as collateral channels of circulation.

Moya moya syndrome - same angiographic appearance associated with conditions such as arteriosclerosis, autoimmune disease, Down syndrome, head trauma, meningitis, neurofibromatosis type 1, and previous radiation therapy.

Bilateral concentric stenosis or occlusion of the **distal internal carotid arteries** and the **proximal anterior and middle cerebral arteries** are consistent pathologic lesions.

Rarely, the posterior cerebral artery or other vessels of posterior circulation are involved.

Diffuse collateral reticulate network of vessels (**moya moya vessels**) - **Hallmark** of the disease.

Etiology:

- Unknown - though myriad bacterial, environmental, genetic, and viral causes have been theorized.
- HLA class II association +
- Familial

Extracranial involvement:

Renal, pulmonary, and coronary vessels.

Diagnosis

Cerebral angiography - Gold standard.

MRI and MRA – other methods of diagnosis but inferior to Cerebral Angiogram.

Cerebrospinal fluid exmn. – Unremarkable

EEG - can be abnormal but are usually *nonspecific*.

Management

No known treatment is curative.

Medical:

- Anticoagulant and antiplatelet agents have shown no remarkable benefit.
- Corticosteroids – not beneficial.

Surgical treatment:

Main modality of Treatment.

- To manage the hemorrhagic and ischemic consequences of moyamoya disease.
- **Revascularization procedures.**
 - Direct
 - Indirect

Direct – usually in adults;

- Superficial temporal artery to middle cerebral artery bypass or
- Middle meningeal artery to middle cerebral artery bypass.

Indirect - traditionally used in children.

Galeo Arachnoid synanastamosis by multiple burr holes.

Encephaloduroarteriosynangiosis (EDAS)

Encephalomyosynangiosis (EMS)

Omental Transposition

Encephalomyoarteriosynangiosis (EMAS)

Increase perfusion in the cerebrovascular territory of the middle cerebral artery.

Do not substantially effect the circulation in the fields of the anterior or posterior cerebral arteries .

Literature review

- Nigel Peter Symss, Ravindranath kapu *et al.* Multiple burr hole surgery as a treatment modality for pediatric moyamoya disease. *J Pediatr Neurosci.* Jul 2010;5(2):115-120.
- 2-yr-old boy presented with recurrent episodes of TIAs with seizures since the age of 6 months – Moya Moya. EDAS done. Now no TIA like episodes. Y.C. Manjunatha and Arun Kumar Gupta. *IJP* - JUL.2010;77(7):817.

Thank You