MOYAMOYA DISEASE

Introduction

- A chronic occlusive cerebro-vascular disease affecting arteries around the ‘circle of Willis’ & formation of extensive collaterals at the base of the brain
- Presents with ischemic and hemorrhagic symptoms
- Characteristic angiographic finding

History

- First described in Japan- Takeuchi & Shimizu (1957)
- Spontaneous occlusion of the ‘ circle of Willis’- Kudo (1968)
- Moyamoya means ‘puff of smoke’
- Coined by Suzuki and Takaku in 1969

Epidemiology

- Highest incidence in Japan (0.35/ lakh)
- Incidence in Western countries- 1/10th of Japan
- F:M= 2:1
- Bimodal age distribution: larger peak in 1st decade & smaller peak around 30-49years
- 10-15% have familial form

Etiology

- Multifactorial: genetic predisposition and environmental stimuli

- Genetic loci: chromosome 3, 6, 8 & 17

Associated conditions

- Immunologic: Grave's disease/ thyrotoxicosis
- Infections: Leptospirosis and tuberculosis
- Hematologic disorders: Aplastic anemia, Fanconi anemia, sickle cell anemia, and lupus anticoagulant

Associated conditions 2

- Congenital syndromes: Apert syndrome, Down syndrome, Marfan syndrome, tuberous sclerosis, Turner syndrome, NF-1& Hirschsprung disease

- Vascular diseases: Atherosclerosis, coarctation of aorta, fibromuscular dysplasia & hypertension

Associated conditions 3

- Others: Head injury, Head neck irradiation for optic glioma, pituitary tumor, craniopharyngioma.

- These are not causative, but warrant consideration during treatment.

Patho-physiology 1

- Smooth muscle hyperplasia of vessel wall & luminal thrombosis

❖Fibro cellular thickening of intima,

❖Attenuation of media

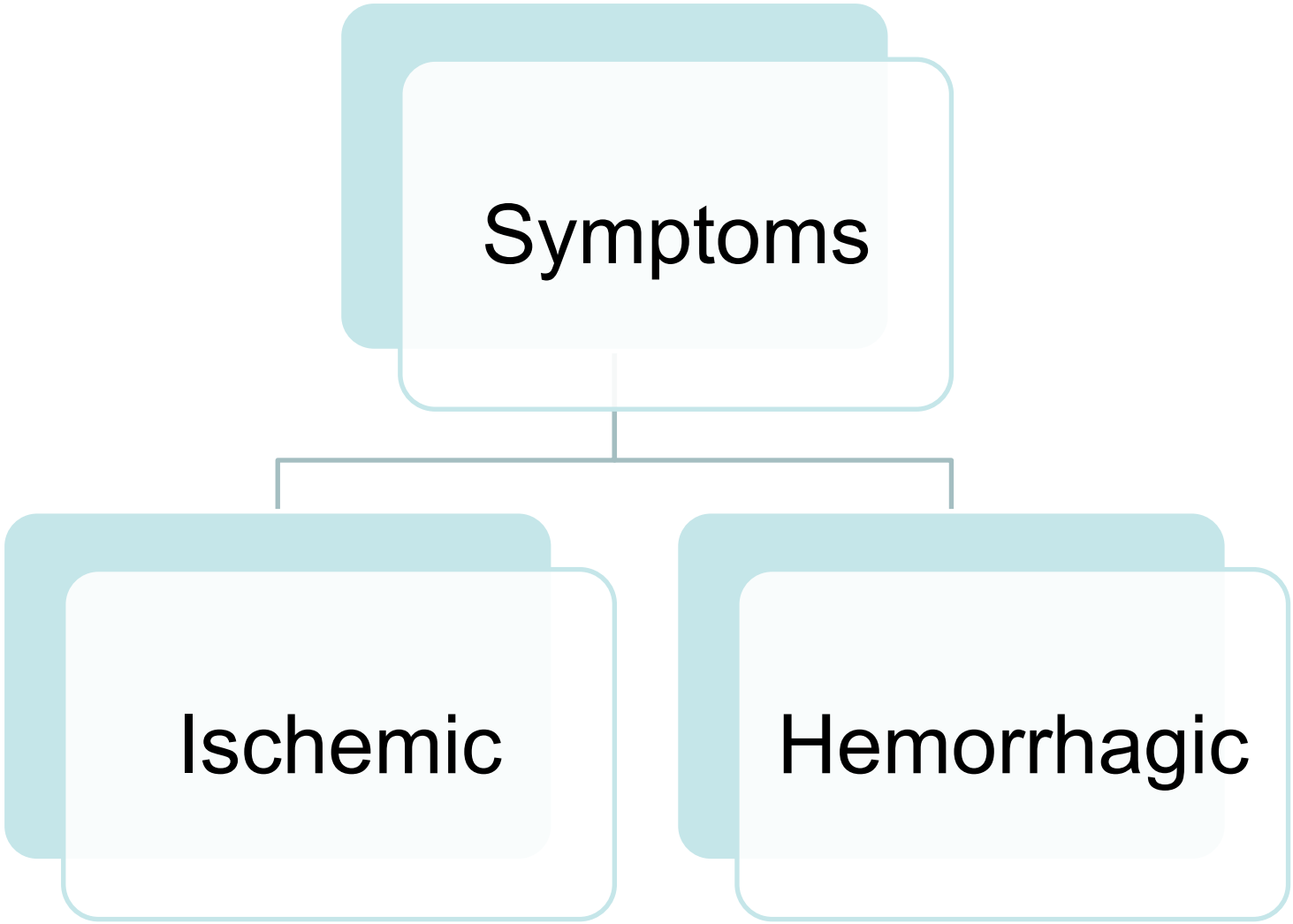
❖Disruption of internal elastic lamina

- No evidence of inflammation or arteriosclerosis

Patho-physiology 2

- Site: supra-clinoidal ICA, ACA & MCA
- Rare involvement of PCA & BA
- Extra-cranial involvement: STA
- Role of pleuripotent peptides, enzymes & receptors: primary or secondary

Clinical features



Pediatric population

- Ischemic symptoms: 70-80% cases
- Stroke or TIA: 6% of childhood strokes
- Occurs in watershed areas
- Precipitating factors:
  - Hyperventilation
  - Dehydration

Pediatric population 2

- Features:

❖Hemi paresis

❖Speech disturbance

❖Cognitive impairment

❖Seizure

❖Subtle deficits: developmental delay, syncope, personality changes, visual disturbance

Pediatric population 3

- Hemorrhage: IVH, intraparenchymal or subarachnoid
- Headache
- Choreiform movements

Adult population

- Hemorrhage: 66% cases
- Intra or periventricular bleeding
- Annual rebleeding rate 7%
- High morbidity & mortality
- Sources:
  - Fragile collateral vessels
  - Micro aneurysms in the circle of Willis
  - Periventricular pseudo aneurysms
  - Saccular aneurysms in vertebro-basilar system

Adult population

- Ischemic symptoms predominate in Western world
- Low morbidity and mortality
- Pregnancy and delivery increase the risk

Imaging

- Angiography: Gold standard
- MRI & MRA: steno-occlusive carotid lesion and basal Moyamoya
- Plain CT: helps in acute stage
- Cerebral blood flow studies: xenon enhanced CT, PET, SPECT

Angiography

- Suzuki & Takaku staging:

❖ Stage 1: Narrowing of carotid fork

❖ Stage 2: Initiation of Moyamoya

Angiography 2

- Stage 3: Intensification of Moyamoya

- Stage 4: Minimization of Moyamoya

Angiography 3

- Stage 5: Reduction of Moyamoya

- Stage 6: Disappearance of Moyamoya

Angiography 2

- Types:

❖Basal Moyamoya

❖Ethmoidal Moyamoya

❖Vault Moyamoya

Management

- No definite treatment available
- Medical treatment: not effective
- Aspirin
- Anticoagulants
- Calcium channel blockers
- Steroids

Surgical management

- Aim:
  - Augment cerebral blood flow
  - Improve cerebral hemodynamics

- Methods:
  - Direct revascularization
  - Indirect revascularization
  - Combined

Surgical management 2

- Criteria for revascularization:

1. Symptomatic patients with good neurological status
2. Infarction <2cm on CT & all previous hemorrhages resolved completely
3. Angiographic stage II to IV
4. Timing: > 2 months after the most recent attack

Direct revascularization

- Indicated when donor & recipient vessel diameter >1mm
- Immediate selective perfusion of ischemic area
- Chance of hyper perfusion syndrome
- Usually done in adults

Direct revascularization 2

- STA-MCA bypass- Donaghy & Yasargil (1967)
- STA-ACA bypass
- STA- PCA bypass

Indirect revascularization

- Aimed at stimulating neovascularization
- Extent of revascularization unpredictable
- Useful in pediatric population

Indirect revascularization 2

- Encephalomyosynangiosis (EMS): implantation of temporalis muscle on lateral brain surface and secured to dura

- Encephaloduroarteriosynangiosis (EDAS): dissected STA is laid onto the cortical surface

Indirect revascularization 3

- Ribbon EDAS: pedicle of galea inserted into interhemispheric fissure
- Autogenic omentum transplantation as free graft

Peri-operative care

- Adequate hydration
- Normo-capnia
- Analgesia
- Normo-thermia

Follow-up

- Clinical evaluation & angiography after 6 months
- Angiography after 1 year
- MRA annually from second year

Assessment of revascularization

- Qualitative:

➢ Matushima grading on DSA-

❖ Grade-A: good revascularization- >2/3rd of MCA territory

❖ Grade-B: fair- 1/3 to 2/3rd of MCA territory

❖ Grade-C: poor- slight or no collateral formation

Assessment of revascularization2

- Qualitative:

➢ Doppler grading after EMS:

❖ Grade 1: no vessel formation

❖ Grade 2: 1-4 vessel formation

❖ Grade 3: >4 vessel formation

Assessment of revascularization3

- Quantitative:
- Study published in Neurosurgery in March 2012
- Quantitative assessment of RV on DSA
- Revascularization of MCA territory against supratentorial area of the ipsilateral hemisphere
- Best result following combined procedure

Prognosis

- Benign course in 75-80%
- Rebleeding occurs in 30-65%
- Revascularization reduces rebleeding & TIAs
- Unilateral disease progresses to bilateral involvement in 7-27%

Future prospects

- Role of endothelial progenitor cells
- Role of cytokines and growth factors
- Quantitative assessment of RV

AIIMS data

- Ten-year experience of 44 patients with Moyamoya disease from a single institution
- Published in Journal of Clinical Neurosciences in April 2010
- Adult population predominates: 59% vs. 41%
- Hemorrhagic symptoms more common: 68% vs. 32%

AIIMS data 2

- Revascularization done in 11 patients: 9 indirect & 2 combined
- No new episode in revascularized patients
- In conservatively managed 19 patients 7 developed new episodes
- In hospital mortality: 3 patients with hemorrhagic symptoms died

Conclusion

- The unpredictable and relentless course of the MMD, coupled with irreversible nature of deficits once present dictates a need for early diagnosis, prompt treatment and regular follow-up


Thank You