AV Malformation

CASE SCENARIO

- Aysha Fathima, 11 yr old, Neurologically normal child
- H/o sudden LOC preceded by headache & 1 episode of vomiting
- No seizures/altered sensorium

O/E : Deep pain responsive

Vitals : Afebrile

Pulse 90/mt

BP 114/70 mm Hg

RR 26/mt

SpO2 – 86%

Both pupils dilated and not reacting to light

GCS : E1 M4 V1 = 6/15

CNS : Left UL and LL weakness

Left facial weakness +

ER MANAGEMENT

- Non convulsive status epilepticus:
  - Antiepileptic measures (Lorazepam, Fosphenytoin & Phenobarbitone was given)
  - Intubated with RSI
  - Increased ICP :
  - 3% Nacl and hyperventilation

MRI BRAIN

Acute ICH in right Centrum Ovale with intraventricular extension of the hemorrhage.

Mass effect in the form of Sulcal effacement and midline shift of 6mm to the left side.

MANAGEMENT

- Neurosurgical management- Dr.Thirumaran
  - Craniotomy done – ICH evacuated
  - 2 midline AVM was removed
  - Compression bandage over craniotomy
  - HPE : consistent with AV Malformation
- Medical :
  - Antibiotics – Ceftriaxone
  - Hypertension – Antihypertensives

CT brain –

cerebral edema decreased

Right SDH with minimal shift to the left

Minimal inter ventricular hemorrhage involving bilateral ventricles

small residual hemorrhage of the right centrum ovale

CSF collection in the right hemicraniotomy site

- Developed subdural collection at craniotomy site – CT done
- Subdural tap was done - compression bandage treatment
- Epidural catheter inserted and continuous CSF drainage done
- CSF analysis – Normal

- Child was shifted to ward
- IV antibiotics, anti hypertensives were continued on discharge.

FOLLOW UP

- At discharge – GCS 15/15, left hemiplegia +, 0 power
- At 6 months –left hemiparesis, no other neurological deficit
- After 1 year – Motor improvement (power 4/5)

  No intellectual deficit

CEREBRAL AVM

- Angio architecture : AVM results from direct arterial to venous connections without an intervening capillary network.
- There is development of afferent and efferent pedicle and arterialization of the venous limb.
- Aneurysms
  - Source of bleeding
  - Worsen the prognosis

EPIDEMIOLOGY

- Brain AVMs occur in 0.1% of the population, one-tenth the incidence of intracranial aneurysms
- Supratentorial lesions – 90%
- Infratentorial lesions – 10%
- 1 to 2 % of strokes, 3% of strokes in young adults, 9% of SAH is due to AVM
- >1 AVM in brain – Osler Rendu Weber syndrome
- Pathogenesis is not well understood

HEMODYNAMICS

- The velocity of blood flow through AVM is higher than through normal brain

- The feeding arteries and draining veins progressively dilate and become tortuous

- They consume large volumes of blood depriving the brain of normal circulation(“steal phenomenon”)

- Space-occupying effect or rupture of a vein and intracerebral bleeding

AGE & LOCATION

- Age - Between 10 and 40 years

- Cerebrum – 60 to 70%
- Cerebellum
- Brainstem
- Spinal cord
- Deep seated

PRESENTATION

- Hemorrhage – commenest symptom, children > adults
- Seizures
- Migraine like headache but on same side
- Focal neurological deficit
- Auscultation of the skull - high-pitched bruit
- Rupture of AVM

  headache, vomiting, nuchal rigidity(SAH)

  hemiparesis, and focal or generalized seizure

USE OF IMAGING

- To establish the diagnosis of AVM
- To make a pre-therapeutic evaluation of AVM to decide about management
- To perform post therapeutic evaluation

DIAGNOSIS

- MRI BRAIN : Location of AVM, in case of hematoma, acute ICH
- MRA : identifies AVMs > 1 cm, inadequate to delineate the morphology of feeding arteries and draining veins
- Cerebral angiography : Gold standard for diagnosis, planning treatment and follow up
- CT BRAIN : Identifies only large AVMs, to rule out bleeds, post treatment evaluation

AVM SYNDROMES

SYNDROMES	COMMON FEATURES	OTHERS
STURGE WEBER SYNDROME	Parietal occipital AVM	Port-wine stain, Macrocephaly, DD/MR, ADHD
OSLER WEBER RENDU SYNDROME	Visceral, mucosal and cerebral AVM	Telengectasia
VON HIPPEL LINDAU	Cerebral, pulmonary AVM	Angiomas, loss of vision – Retinal detachment, vitreous hemorrhage

TREATMENT

Medical

- Antiepileptic Rx
- Analgesics

Surgery

- Microsurgery - MC Rx
- Radiotactic surgery
- Embolization

ASA GUIDLINES

SURGERY

- SURGICAL EXTIRPATION
  - Low grade lesion (I and II)

- RADIOSURGERY
  - small lesions associated with increased risk based on anatomy and feeding vessels

- Endovascular embolization then surgery
  - grade III lesions

- CONSERVATIVE
- Grade IV,V

PROGNOSIS

- AVM hemorrhage is the strongest predictor of rebleeding in untreated brain AVM, but less in children when compared to adults.
- Prognosis after AVM hemorrhage is better than after ICH

CARRY HOME MESSAGE

- Any child presenting with seizures/ headache or sudden LOC, AVM should always be considered as a differential diagnosis.

- MRI is the diagnostic investigation of choice for AVM

- Always work in tandem with neurosurgeon for the best outcome

Thanks to

- Dr Thirumaran - Neurosurgeon
- Dr Mahesh - Neurologist
- PICU – Dr Anitha and team