A CASE OF PIAL ARTERIO-VENOUS MALFORMATION
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PRESENTATION OF CASE
A 68-year-old female housewife presented with complaints-
- a. Seizures since 30 years on and off
- b. Headache since 30 years left sided parieto-temporal

Patient has been on treatment since 30 years when she had first episode of seizure, but was not relieved of seizures.

Patient was on following anti-epileptics-
1. Phenytoin 300 mg OD
2. Carbamazepine 400 mg TDS
3. Sodium Valproate 750 mg OD
4. Clobazam 10 mg TDS.

Patient had been taking treatment from various hospitals but was never free of symptoms. Multiple CT (computerized tomography) scans, MRI (magnetic resonance imaging) studies had been done but no definitive diagnosis was achieved.

Patient used to have seizure episodes lasting from 5 minutes to 15 minutes, once or twice a day, associated with tongue bite, frothing from mouth, up rolling of eyeballs and loss of consciousness from 15 minutes to 1 hour.

Headache spells were associated with nausea sometimes vomiting.

When patient was brought to Dr. D.Y. Patil Hospital, she had an episode of seizure at her home and was brought to emergency room in unconscious state. Patient was stabilized with loading dose of Levetiracetam 1 gm in Infusion.

Emergency CT was done which was S/O Gliotic Area in left frontal lobe with small foci of cortical calcification.

Figure 1. CT Brain Showing Gliotic Area in Left Frontal Lobe with Small Foci of Cortical Calcification
Patient was then admitted to wards after stabilization and was started on following medications:
Levetiracetam 500 mg BD
Phenytoin 100 mg TDS
Clobazam 10 mg TDS.

Patient had no convulsions/seizures after admission, hence after stabilization, patient was advised MRI Brain with Contrast and Angiogram.

Figure 2. MRI Brain Showing AV Malformation of Left Frontal Lobe
Figure 3. MRI Brain Showing AV Malformation of Left Frontal Lobe

Figure 4. MRI Brain Showing AV Malformation of Left Frontal Lobe
Report was S/O a well-defined bunch of flow voids involving the left frontal lobe measuring approximately 3.5 x 3 cms. Mild adjacent gliosis is noted.

Prominent feeding arteries are seen arising from left Middle Cerebral Artery supplying the lesion with draining vessels noted draining into the Superior Sagittal Sinus.
**DIFFERENTIAL DIAGNOSIS**
- Cavernous Sinus Syndromes
- Cerebral Amyloid Angiopathy
- Cerebral Aneurysms
- Cerebral Venous Thrombosis
- Dissection Syndromes
- Intracranial Haemorrhage
- Moyamoya Disease.

**CLINICAL DIAGNOSIS**
On the basis of history and clinical presentation, my patient was a case of Seizure Disorder with Migraine.

**DISCUSSION**
Cerebral AV-Malformations (CAVMs) are the most common symptomatic vascular malformations. Possible presentations include:
- Incidental finding in asymptomatic patients: 15%
- Seizures: 20%
- Headaches
- Ischemic events due to vascular steal from normal brain
- Haemorrhage: 65% 5, 2-3% per year:
  - Parenchymal
  - Subarachnoid
  - Intraventricular

A brain arteriovenous malformation may not cause any signs or symptoms until the AVM ruptures, resulting in bleeding in the brain (haemorrhage). In about half of all brain AVMs, haemorrhage is the first sign. But some people with brain AVM may experience signs and symptoms other than bleeding related to the AVM.¹

The origin of arteriovenous malformations remains uncertain, although they are thought to be congenital and perhaps involves dysregulation of vascular endothelium growth factor (VEGF).

AVMs comprise of a number of components
- Feeding arteries
- Nidus
  - Shunting Arterioles: The true culprit
  - Interconnected Venous Loops
- Draining veins.²

The nidus is fed by one or more arteries and drained by one or more veins. The feeding arteries are enlarged due to the increased flow and flow related arterial aneurysms are encountered.

Venous aneurysms also referred as venous pouches, are also seen. It may contain dystrophic calcification, a small amount of gliotic tissue and blood in different stages of evolution.³

**DISCUSSION OF MANAGEMENT**

Angiography (DSA) - Remains the gold standard, able to exquisitely delineate the location and number of feeding vessels and the pattern of drainage. Ideally, angiography is performed in a biplane system with a high rate of acquisition, as the shunts can be very rapid.

On angiogram, AVM appears as a tightly packed mass of enlarged feeding arteries that supply central nidus.

One or more dilated veins drain the nidus and there is abnormal opacification of veins in arterial phase (early venous drainage), representing shunting.⁴ Medications also may be used to treat symptoms caused by the AVM, such as headaches or seizures.

Surgery is the most common treatment for brain AVMs. There are three different surgical options for treating AVMs:

1. **Surgical Removal (Resection).**
   If the brain AVM has bled or is in an area that can easily be reached, surgical removal of the AVM via conventional brain surgery may be recommended. In this procedure, Neurosurgeon removes part of the skull temporarily to gain access to the AVM.
   With the help of a high-powered microscope, the surgeon seals off the AVM with special clips and carefully removes it from surrounding brain tissue. The surgeon then reattaches the skull bone and closes the incision in the scalp.
   Resection is usually done when the AVM can be removed with little risk of haemorrhage or seizures. AVMs that are in deep brain regions carry a higher risk of complications. In these cases, other treatments are recommended.⁵

2. **Endovascular Embolization**
   In this procedure, surgeon inserts a catheter into a leg artery and threads it through blood vessels to the brain using X-ray imaging. The catheter is positioned in one of the feeding arteries to the AVM, and injects an embolising agent, such as small particles, a glue-like substance, microcoils or other materials, to block the artery and reduce blood flow into the AVM. Endovascular embolization is less invasive than traditional surgery. It may be performed alone, but is frequently used prior to other surgical treatments to make the procedure safer by reducing the size of the AVM or the likelihood of bleeding. In some large brain AVMs, endovascular embolization may be used to reduce stroke-like symptoms by redirecting blood back to normal brain tissue.⁶

3. **Stereotactic Radiosurgery (SRS)**
   This treatment uses precisely focused radiation to destroy the AVM. It is not surgery in the literal sense because there is no incision. Instead, SRS directs many highly targeted radiation beams at the AVM to damage the blood vessels and cause scarring. The scarred AVM blood vessels then slowly clot off in one to three years following treatment.
   This treatment is most appropriate for small AVMs that are difficult to remove with conventional surgery and for those that haven’t caused a life-threatening haemorrhage.⁷

**FINAL DIAGNOSIS**
From this MRI report and after thorough discussion with radiologists, we came to a conclusion that the MRI findings were suggestive of Pial Arterio-Venous Malformation.
Management of the Patient - Neurosurgical Intervention was required for further management, hence a Neurosurgery reference was taken.

After examining the patient and the MRI findings Neurosurgeon advised the following treatment options to the patient:
1. Surgery after Temporary Therapeutic Embolization, but the risk of Right Sided Hemiplegia and Aphasia had to be kept in mind.
2. Permanent Embolization by Neuro Interventional Radiologists at KEM Hospital.
3. Gamma Knife Procedure at Hinduja Hospital.
4. Life Time Anticoagulant Therapy along with one of the above procedures.

Patient after understanding the risks and poor socio-economic status underwent Surgery after Temporary Embolization at KEM hospital, some hours after surgery patient developed weakness of right side of the body and Motor Aphasia which was the risk of the surgery. But patient was discharged on anti-coagulants and ongoing anti-epileptics. Patient is bed ridden since then.

REFERENCES