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## CASE REPORT

# Arterio-Venous Malformation- A Case Report

**Jahan Afroze[1], Musavvir Samin[2], Sadia Jahan Boni[3]**

**Abstract:**

*An adult female with sudden attack of 2 episodes of seizures came to the hospital semiconscious. NECT was done and diagnosed as ICH-frontal lobe. Subsequently MRI and MRA were done and differentials were Cavernoma, AVM and Neoplastic lesion. Conservative treatment was given and patient was advised for DSA after 4 weeks. After DSA the lesion was diagnosed as AVM. Surgically the lesion was removed and histopathology confirmed the diagnosis.*

*Key Words: AVM, Cavernoma, Hemorrhage, DSA, CTA, MRA.*

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**Introduction:**

A brain Arteriovenous Malformation (AVM) is a tangle of abnormal blood vessels in the brain or on the surface bypasses normal brain tissue diverts direct blood from the arteries to the veins. Brain AVMs occur less than 1% of the general population. It is estimated that about one in 200000 people may have AVM. The arteries are responsible for carrying oxygenated blood for the metabolic demands of the brain and the veins carry deoxygenated blood from the brain to the heart while AVM disrupting this vital pattern.

**Case report:**

A young physician of 25 years came to the emergency room with history of fever and convulsion of 2 times. Patient was febrile for last 3 days and fever was low grade and continuous in nature, not associated with chill and rigor or photophobia. Later she had 2 episodes of generalized convulsion. She also had loss of history of brief period of loss of consciousness during convulsion. She had no history of such episode previously. No history of trauma or limb weakness. General and systemic examination revealed no abnormality except for right sided grade 1 facial weakness. Her routine hematological and biochemical reports were within normal limit. The CT brain showed left frontal ICSOL with tumor hemorrhage with perilesional edema. MRI with contrast was done and showed blood in the left frontal area didn't have significant contrast enhancement. She was put on anticonvulsant. She had no seizure during the hospital stay and she was afebrile. She was discharged with an advice to do Digital Subtraction Angiography 1 month later to confirm the diagnosis. DSA was done and a cortical AVM was found in the left frontal area, flow of which was moderate. The flow came from M4 branch and draining vein went through bridging vein SSS. The size of the AVM was approximately 2*2 cm. there was no flow related or intra nidal aneurysm but there was long subarachnoid venous course. The impression was Spetzler and Martin grade 1 AVM in left frontal cortex. A left frontal craniotomy and total excision of the arteriovenous malformation was done. She had satisfactory postoperative period without recruitment of any new neurological deficits. She had no seizure, fever or weakness of any part of the body. She was discharged with the advice of doing another DSA three months later.

1. Assistant Professor, NICVD, Dhaka
2. Honorary Medical Officer, SSMC & MH, Dhaka
3. Honorary Medical Officer, SSMC & MH, Dhaka

**Correspondence to:** Dr. Jahan Afroze, Assistant Professor, NICVD, Dhaka, Mobile: 01711-861950, E-mail: jahanssmc10@gmail.com

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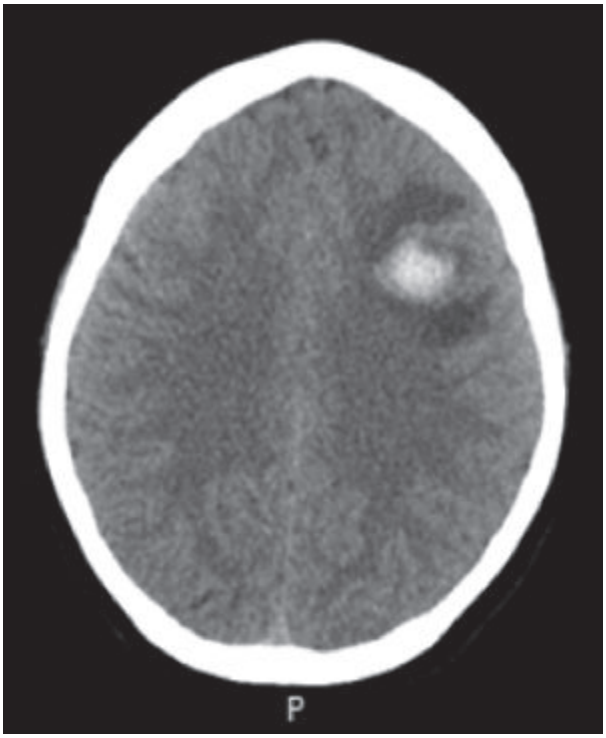
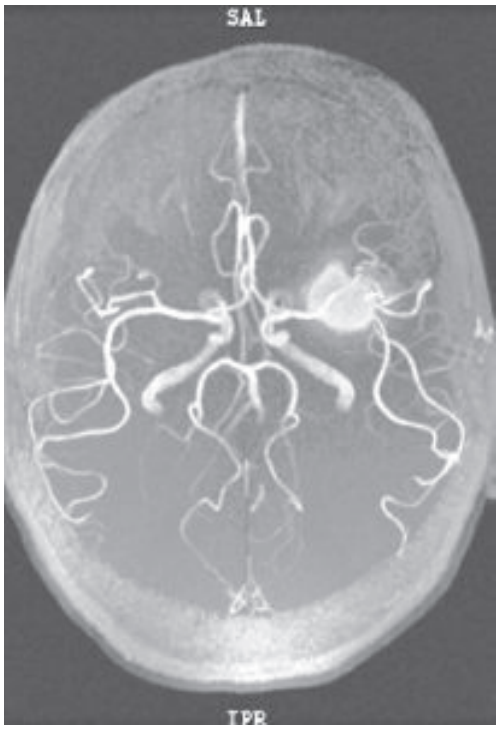
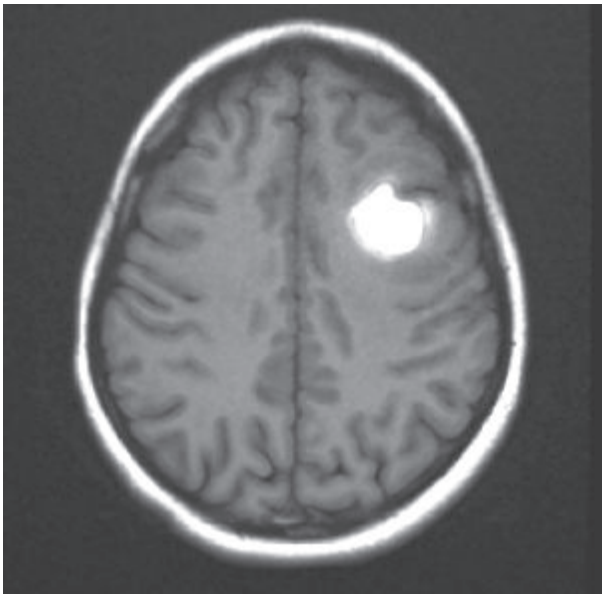
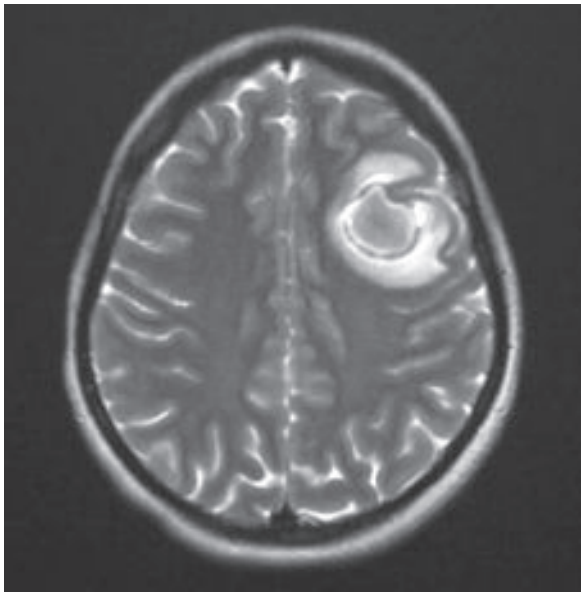



NECT scan of Brain showing Hyperdense lesion with Perilesional oedema

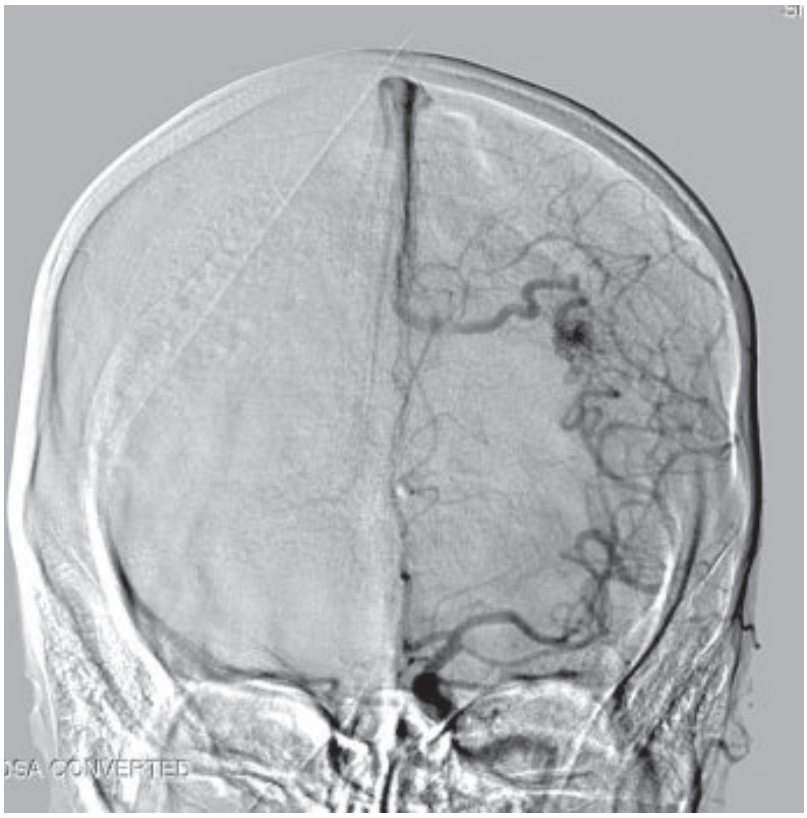



MRA showing mid segmental proximal left MCA is not well visualized and pushed by the ICH in left frontal lobe with crowding of aberrant vessels in the vicinity of ICH and Prominent vein draining into Superior Sagittal Sinus.

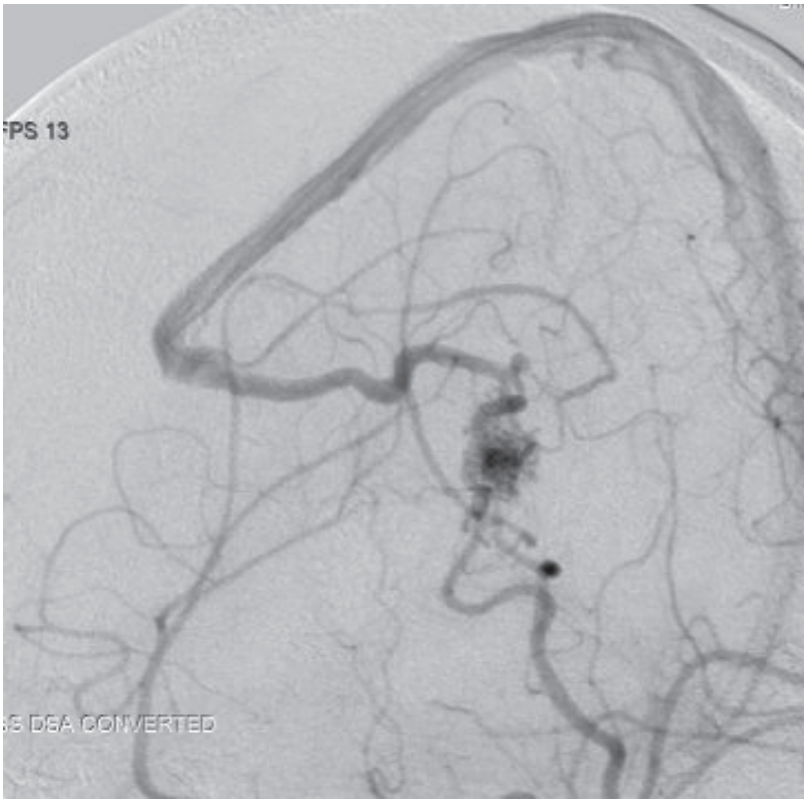




MRI showing Mixed Signal Intensity Lesion having hypointense core bordered by hyperintense rim in T1WI and hypotense in T2WI with Perilesional oedema and compression of the ipsilateral sulci.



DSA showing cortical AVM in Left Frontal Lobe



DSA showing cortical AVM in Left Frontal Lobe

**Discussion:**

AVM stands for Arteriovenous Malformation. An AVM is a tangle of abnormal and poorly formed blood vessels (arteries and veins). They have a higher rate of bleeding than normal vessels. AVMs can occur anywhere in the body. Brain AVMs are of special concern because of the damage they cause when they bleed.

AVM arises about 3 weeks after conception at the time when blood vessels are dividing into vein and arteries.[1] Recent studies have suggested AVMs are dynamics and have the ability to grow, regress and regenerate following obliteration by surgery or radiosurgery. Currently it is thought that the altered expression of more than 900 genes are involved in the pathogenesis of AVM.[2] AVM have been associated with aneurysm in 10-15% of patient group depending of the type of angiographic techniques employed.[3]

Vascular malformations are of 2 types.

A. Low flow varieties LFV: capillary, venous and lymphatic malformation

B. High flow varieties HFV: AVMs and arteriovenous fistula

Location of AVM: (5)

- Supratentoirial 85%
- Infratentorial 15%

Incidence:

- Solitary AVM 98%
- Multiple 2%

Associated abnormalities-

- Flow related angiopathy secondary to endothelial hyperplasia
- Flow related aneurysm:
  - Intranidal
  - Intrapedicular
- Remote aneurysm: Hemodynamically unrelated to aneurysm

Classification:

1. Compact (Glomerular) Nidus: Abnormal vessels without any interposed normal brain tissue.
2. Diffuse (Proliferative) Nidus: No well-formed nidus is present with functional neuronal tissue interposed amongst the anomalous vessels.

Basic pathological characteristics of AVM: it demonstrates variable histology that may reveal capillaries, venules and arterioles that may exhibit hypertrophy, wall thickening and possibly dystrophic calcification all within a densely fibrous or fibromyxomatous background. [10]

The patient of an AVM at which rapid arterial shunting passes into an arterialized vein acting as a low pressure cell is termed the nidus.[16] The increased shunting into the nidus lead to arterial and venous hypertrophy that can compress erode infiltrate or destroy adjacent normal structures.[17,18]

Spetzler Martin AVM grading system: It is a method of estimating the mortality and morbidity of surgical resection to guide treatment recommendation. It allocates points for various features of intracranial AVM including size, eloquent location and venous drainage.[23]

Size of the nidus

| | |
|---|---|
| 1 point | Small < 3 cm |
| 2 point | Medium 3-6 cm |
| 3 point | Large > 6cm |

Eloquence of adjacent brain:

| | |
|---|---|
| 0 point | Non eloquent ( frontal and temporal lobe, cerebral hemisphere) |
| 1 point | Eloquent ( sensory, motor, language, visual cortex, hypothalamus, thalamus, bnrain stem, cerebellar nuclei or region directly adjacent to these structures |

Venous drainage:

| | |
|---|---|
| 0 point | Superficial only |
| 1 point | Deep |

The Spetzler Martin Grading was originally validated in a study of 100 consecutive patients treated with microsurgical excision of AVMs. Morbidity rates were as follows:

| Grade | Morbidity % |
|---|---|
| 1 | 0 |
| 2 | 2.5% minor deficit |
| 3 | 12% minor deficit, 4% major deficit |
| 4 | 20% minor deficit, 7% major deficit |
| 5 | 19% minor deficit, 12% major deficit |
| 6 | Used to describe inoperable lesion |

The score correlates with operative outcome.

Investigation: The first imaging study that is performed in patients with a suspected AVM is usually a computed tomography (CT) or magnetic resonance imaging (MRI) scan. These studies are good for depicting AVMs and they are relatively noninvasive, only requiring an injection of contrast material into a small vein. Overall, arterio venous malformations are best imaged by using MRI, which can uniquely show these lesions as a tangle of vascular channels that appear as flow voids. Nonenhanced CT is superior for visualizing the small foci of calcification often associated with arteriovenous malformations, and it may also delineate hyper attenuating serpiginious vessels that constitute the nidus.

Non enhanced CT scan is valuable for demonstrating the extent of acute hemorrhage and hydrocephalus. Contrast-enhanced CT shows enhancement of the typical vascular channels. Magnetic resonance angiography (MRA) or CT angiography (CTA) may be adequate for initial or follow-up evaluation of an arterio venous malformation.

CT scan is an excellent examination for detecting cerebral hemorrhage, but it can miss an underlying AVM. AVMs are typically iso attenuating relative to normal parenchyma and, therefore, can be overlooked, particularly if a contrast agent is not administered. In an emergency setting, the administration of an iodinated contrast agent is typically deferred in favor of patient stabilization. Contrast-enhanced CT scanning also poses an inherent risk of radiation and, because of its cost, MRI may be a better screening examination for AVM in the general population. Contrast-enhanced CT scanning is performed to detect cerebral AVM, however, when MRI is contraindicated or otherwise not feasible.

MRI is excellent for demonstrating the AVM nidus and abnormal flow voids typical of an AVM; however, in acute cerebral hemorrhage, compressed AVMs may no longer demonstrate flow and may, therefore, be overlooked. This may lead to the need for serial MRI studies to search for an underlying cause of cerebral hemorrhage not shown on a single MRI study. MRI can cause underestimation of the number of feeding arteries and associated aneurysms, which might also be missed. Furthermore, MRI can have a relatively poor sensitivity in detecting dural malformations. Gadolinium-based contrast material may be needed to demonstrate abnormal vascular channels.

Digital substraction angiography (DSA) remains the criterion standard for characterization and delineation of brain and spinal AVMs. Angiography is a dynamic real-time study that not only demonstrates the presence or absence of an AVM, but also shows vascular transit time. Diagnostic angiography is uniquely able to delineate the size and number of feeding arteries, and it can define the pial, dural, or mixed origin of the AVM.

Angiography can be used to measure the size of the AVM and judge the compactness of the nidus. Furthermore, angiography can be used to evaluate the venous drainage pattern (superficial, deep, or mixed). In addition, angiography frequently depicts associated risk factors for hemorrhage, including aneurysms and venous stenosis. Planning an angiography is a vital step in both interventional neuroradiologic and neurosurgical evaluation of patients with AVM. [11]

Angiography can reveal certain features that are believed to correlate with an increased risk of hemorrhage. These features include the presence of associated intranidal, remote, or pedicular aneurysms; central or deep venous drainage; stenosis of a draining vein; and a periventricular or intraventricular location.

Conclusion: DSA remain the gold standard for the diagnosis of intracranial AVM but is invasive. CT scan easily identifies intracerebral hemorrhages, raising suspicion of AVM in a younger person or a patient without clear risk factors for hemorrhage; however, this modality can identify only large AVMs with the disadvantage of radiation whereas existing MRA is suboptimal for assessing the hemodynamic information with a temporal resolution of 50-100 for the evaluation of AVMs and can complement existing methods such as DSA, TOF MRA, DMRA, and BDCT angiography.

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