Cerebrovascular Malformations with Dural Sinus Thrombosis with Extensive Parenchymal Calcification

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INTRODUCTION

Cerebral vascular malformations refer to an abnormal connection between a vein and an artery. Although no specific cause has been identified, a connection to CVT (Cerebral Venous Thrombosis) has been posited. We are demonstrating cerebral vascular malformations with cerebral venous thrombosis according to dynamic changes in the venous drainage pattern and intracranial pressure. This observation insights into the pathophysiological association between cerebral vascular malformations with CVT.

We report a case of dural AVF (Arterio-Venous Fistula) and pial AVM (Arterio-Venous Malformation) with B / L transverse sinus thrombosis. BAVMs (Brain Arterio-Venous Malformation) can be classified according to their blood supply - "Pial AVM are within the brain parenchyma and are fed by ICA (Internal Carotid Artery) or vertebral artery." Dural AVM are uncommon, and most of the times are infratentorial and considered to be sequelae to surgery, accidents, thrombosis of an or veno-occlusive disease or adjacent venous sinus. They receive blood from ECA (External Carotid Artery) and drain into the sigmoid and transverse sinuses, however these might sometimes include the cavernous venous sinus, superior sagittal sinus, inferior petrosal sinus or areas other than these of the venous system of the brain.[1] "Mixed AVMs mostly when it has supply from both ICA and ECA.[2]

DAVFs (Dural Arterio-Venous Fistula) are a variable group of anomalies that share AV (Arterio-Venous) shunts from dural vessels. They can appear on imaging with haemorrhage or venous hypertension which makes them difficult to manage. Definite conclusion is mostly difficult on NCCT but can be thought of if an intracranial haemorrhage is not in a definitive arterial territory or age category. With contrast enhanced angiography a definitive conclusion is made easily most of the times. Findings that suggest including abnormally tortuous and enlarged vascular channels in the subarachnoid space, these are in fact dilated cortical veins, an enlarged ECA or expanded trans osseous channels and any unusual venous sinuses together with arterialization of contrast phase seen within the unusual sinus attributed to AV shunt formation. In subjects with reverse venous drainage, oedema can be seen in almost 1 / 2 the subjects, even though this can also be presented in subjects who do not have reverse drainage on contrast enhanced angiographic investigation. The locations where white matter oedema is present can show enhancement. These detections are suggestive of a hostile DAVF which has a huge risk of haemorrhage.[3]

BAVMs do not include pure Galen vein AVMs, dural arteriovenous fistulas, cavernous malformations, venous malformations, venous varices or any of the other rarer types of cerebrovascular anomalies.[4] Of BAVMs, 82 % are lobar and 12 % are infratentorial while 9 % of all are deep. 70 % of supratentorial AVMs are purely pial while approximately 50 % of posterior fossa AVMs are purely pial, the rest being purely dural or mixed pial-dural.[5] 48 % generally are seen at watershed locations (involving more than one arterial territory).

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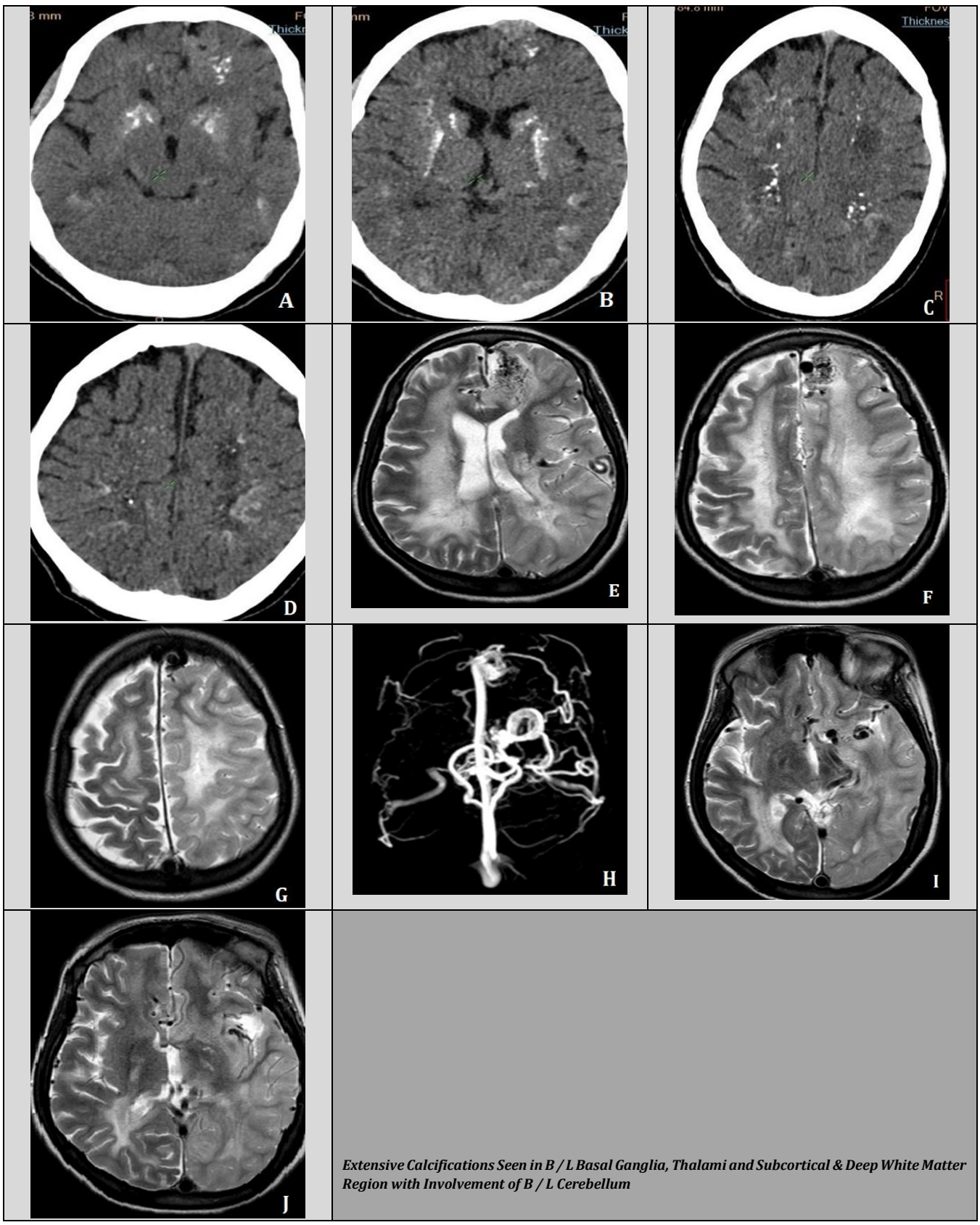
DOI: 10.14260/jemds/2020/807

How to Cite This Article:
Gakhar A, Madaan U, Gupta R, et al. Cerebrovascular malformations with dural sinus thrombosis with extensive parenchymal calcification. J Evolution Med Dent Sci 2020;9(48):3683-3686, DOI: 10.14260/jemds/2020/807

Submission 24-08-2020,
Peer Review 17-10-2020,
Acceptance 24-10-2020,
Published 30-11-2020.

PRESENTATION OF CASE

A 30-year-old woman came with abnormal body movements, altered sensorium with recurrent episodes of seizure since 1 month. She was bed ridden since then however, her condition worsened since 4 - 5 days. She was treated with clobazam and phenytoin. She had past history of type II diabetes myelitis.



Extensive Calcifications Seen in B / L Basal Ganglia, Thalami and Subcortical & Deep White Matter Region with Involvement of B / L Cerebellum

During her stay in the hospital, she mentioned that the seizures as well as sensorium had improved gradually. Non contrast CT scan of the head was done. (Image A - D) and it shows extensive calcifications in B / L basal ganglia, thalami and subcortical & deep white matter region with involvement of B/L cerebellum. Focal clustered calcifications were also seen in left frontal regions. Initially diagnosis of metabolic disorder like Fahr's disease and hyperparathyroidism was considered. MRI (Magnetic Resonance Imaging) was advised for further evaluation.

Tuft of closely placed multiple abnormal flow voids / vessels were seen in left frontal regions (Images E, F) in parafalcine location without any interposed normal brain parenchyma with nidus measuring approximately 2.7 X 2 X 2.6 cm consistent with pial AVM. Few foci of haemorrhage was seen on FFE (Fast Field Echo) sequence. Arterial feeders were seen from left ACA (Anterior Cerebral artery). Few tiny intra lesional aneurysm were seen measuring up to 2.5 mm.

Extensive gyriform areas of signal abnormality appearing hyperintense on T2 / FLAIR (Fluid-Attenuated Inversion Recovery) / DWI (Diffusion-Weighted Imaging) images with mild restriction on ADC mapping were seen involving the cortex in almost whole of the left cerebral hemisphere and left thalamus with extensive T2 / FLAIR hyperintensities were seen in B/L cerebral periventricular and subcortical white matter more marked in B / l fronto-parietal regions. (Image F & G)

On venography there was suggestion of dural AV fistulas with multiple dilated and tortuous flow voids seen in left cavernous sinus region and in left perisylvian region on venography images B / L transverse sinuses show no signal (Image H) written informed consent was obtained from the patient for publication.

DISCUSSION

Lack of oxygen leading to new vessel formation or a reopening of previously present vascular channels by venous hypertension are thought of as the cause of dural arteriovenous fistula and pial AVM with CVT. In our case, there was a possible shift of the drainage to the venous system of the brain, which most likely caused continuing cortical venous hypertension. This venous hypertension must have led to the formation of dAVFs and pial AVM and cortical brain calcification.

Characteristic	Number of Points Assigned
Size of AVM	1 Point
Small (< 3 cm)	2 Point
Large (> 6 cm)	3 Point
Location	
Noneloquent Site	0 Point
Eloquent Site*	1 Point
Pattern of Venous Drainage	
Superficial Only	0 Points
Deep Component	1 Point
Sensorimotor, Language, Visual Cortex, Hypothalamus, Thalamus, Internal Capsule, Brain Stem, Cerebellar Peduncles, or Cerebellar Nuclei	

BAVMs is also microscopic or large enough to involve a complete hemisphere. BAVM volume has been characterised by the dimensions of the nidus, into four groups: huge (> 50 mL), large (25 - 50 mL), moderate (10 - 24 mL), small (< 10 mL). Diameters are measured within the middle to late arterial phase of the contrast enhanced angiography, with check on magnification. Most AVMs are small (< 3 cm, 87 %).[6]

Pial AVM has been graded per Spetzler-Martin grading system. Based on this method[7] the chance of surgery is estimated, which consists of three elements: size, venous drainage, and site.

BAVMs are haemodynamically compartmentalized; each of them possesses feeding arterial channels and draining veins. The number of domains in an AV malformation depends on its dimensions. An AV malformation < 3 cm in size is most likely going to own 1 compartment, a slightly bigger AV malformation (3 - 4 cm) may have 2 of them, and larger AV malformation (> 4 cm) in size most likely has a minimum of 3 compartments. Blood passing within an AV malformation is in direct correlation with the number of compartments and to AV malformation dimensions.

Diffuse AV malformations are disseminated within the brain parenchyma and are generally seen within the deep grey matter. Regional blood flowing around an AV malformation could also be reduced to 81 % of normal referral because of the steal phenomenon the entire cerebral blood flow is also increased by the maximum amount as 50 - 100 %.

Due to ischemia, the neighbouring parenchyma undergoes atrophy and gliosis and discoloured by haemosiderin after prior haemorrhage, with production of scattered foci of calcification, because of the slow progressive growth.

MRI can demonstrate areas of AVM involvement with its size and shows both the dilated feeding arteries and enlarged draining veins. The accompanying dilatations on arterial feeders and accompanying outcome like mass effect, oedema, or ischemic changes also are discernible. BAVMs are difficult to determine on CT (Computed Tomography) but large haemorrhage may be evaluated by MR. MRI is especially compatible to document AVM rupture. However, the ability of MRI (Magnetic Resonance Imaging) to detect aneurysmal dilatations < 1 - 2 cm is less. Those lesions which are not seen on contrast enhanced CT angiography could also be found on MRIs only because they can detect haemosiderin deposits or other proof of disintegration of blood. MRA (Magnetic Resonance Angiography) may be a non-invasive alternative to standard angiography. 3D dynamic MRI with multiple surface coils and parallel images is the best used yet with least interobserver discrepancy and depiction of direction, rate, and quantity of blood flow which is particularly important if embolotherapy is planned.

DISCUSSION OF MANAGEMENT

The management of intracranial AV malformations generally includes embolization, direct surgical or microsurgical resection or radiosurgery. Mostly tiny or insignificant AV malformations located in non-eloquent locations of the brain will be taken out following conventional microsurgery. However, bigger AV malformations in critical locations generally need proper staging and multistep nursing and therapy. Embolization can be done alone when invasive procedures are not prescribed or subject is not willing or will be done before surgery to scale back the degree of the AVM

nidus or may follow microsurgery, radiosurgery, or both. Various agents are used for embolization.[8]

Financial or other competing interests: None.
Disclosure forms provided by the authors are available with the full text of this article at jemds.com.

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