Spectrum of Imaging Findings in Cerebral Venous ischemia

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Learning objectives

Ischemia is defined as a decrease in blood supply to an organ caused due to obstruction of the blood vessels. The definition of venous ischemia is not established. Venous ischaemia can be diagnosed by prolonged circulation time & dilatation with tortuosity of the veins on cerebral angiography. However, the clinical features of venous ischaemia vary based on the underlying etiology, degree of collateral venous compensation, age at onset, duration of onset & the severity.

We propose to describe the underlying etiology, classify & with the help of diagrams & images illustrate the role of cerebral venous ischemia (CVI).

Background

Identification of patients with cerebral venous ischemia is difficult due to lack of any telltale signs of venous insufficiency. Though, venous vascular territories are well described, the diagnosis of CVI is often made by exclusion of arterial ischemia in adults. In children, there is a much lower threshold in the suspicion for CVI.

Based on the underlying pathophysiological mechanism, venous ischemia can be classified as due to:

1. Thrombosis of cortical vein or dural sinus
2. High grade stenosis of major dural sinuses
3. High flow arteriovenous shunting with impaired venous outflow of the cerebral, spinal cord or orbital parenchyma
4. "Secondary Pseudotumor Syndrome" and
5. Hydrodynamic disorders due to CVI in children (Figure 1).

1. Normal Anatomy: (Figure 1a)

The cerebral venous system consists of the superficial, the deep venous systems & the infratentorial venous system.

The superficial venous system is formed by multiple small cortical veins draining into the adjacent dural venous sinuses. The dural venous sinuses drain in anterior to posterior direction with the exception of the superior petrosal sinus which drains in a posterior to anterior direction into the cavernous sinuses. These dural sinuses are large low pressure veins attached to the duramater which leave the skull base as right & left jugular venous sinuses & through multiple emissary veins at the base of skull into the condylar & suboccipital venous plexuses.
The deep venous system is formed by multiple small medullary, subependymal veins & by a few cortical veins which drain into the internal cerebral vein & the vein of Galen. The deep venous system also drains in an anterior to posterior direction.

The infratentorial venous system is a complex system of cortical veins & internal parenchymal veins which drain with no particular directional sense. They generally drain into large veins & sinuses that are located in their immediate vicinity.

The normal venous system & some of the pathologies associated with CVI are illustrated in Figure 1.

B. Venous Territories: (Figure 2)

According to Medler et al. the venous territories though varied, have a definitive drainage pattern as illustrated. The infratentorial venous system is more complex & it cannot be characterised by a straightforward pattern approach.

C. Pathophysiological Mechanism of CVI: (Figure 3a)

CVI results in venous insufficiency of the cerebral parenchyma. The parenchymal abnormalities in CVI are varied & consist of:

1. Initial stage with normal parenchyma but perfusion abnormalities in the form of decreased rCBF and increased rCBV.
2. Vasogenic edema
3. Cytotoxic edema
4. Hemorrhage & hemorrhagic infarction

Unlike arterial ischemia, parenchymal edema due to CVI is reversible.

D. Special Notes:

a. CVI in children as "Hydrodynamic Disorder". (Figure 3b)

CVI in fetus, neonates & infants manifest as "Hydrodynamic Disorder". In neonates & infants, both the interstitial fluid (ISF) & CSF are absorbed by the medullary veins & cerebellum. The separation of the ISF & CSF resorption circuits occurs with cavernous capture of the sylvian veins. Prior to cavernous rupture, the entire venous system has a single outlet through the jugular veins. After the capture, the brain has alternate pathways...
hrough the orbit, pterygoid venous plexuses & the inferior petrosal sinuses. In the presence of "high flow shunts" prior to the capture the maturation of arachnoid villi in the posterior 2/3rds of superior sagittal sinuses gets delayed, impeding CSF absorption. The only remaining drainage through the medullary veins & cerebellum gets decompensated, resulting in retention of both the ISF & CSF.

This results in the typical manifestations of hydrodynamic disorder namely,

1. Cranial vault thickening
2. Macrocrania
3. Supratentorial hydrocephalus
4. "Slit-like" 4th ventricle
5. Tonsillar herniation and
6. "Melting Brain Syndrome" due to abrupt hydrodynamic disturbance & sutural closure causing rapid cerebral atrophy.

b. CVI due to "Secondary Pseudotumor Syndrome". (Figure 3c & 3d)

"Secondary Pseudotumor Syndrome" due to venous sinus thrombosis occurs in 20 to 40% of all patients. These patients present with severe headache, papilledema & progressive visual loss. Also, this can occur due to bilateral transverse sinus stenoses. These stenoses are classically present in the lateral 1/3rd of the transverse sinuses. The imaging features include a partially empty sella", tortuosity of the otic nerve sheaths, increased fluid in the sheaths, flattening of the posterior sclera & "slit-like" ventricles.

E. Classification of CVI based on morphology: (Figure 4)

We would like to propose a classification fo CVI based on amount of brain parenchyma affected.

Focal CVI - Restricted to grey or white matter adjacent to an isolated cortical or medullary vein

Local CVI - Restricted to a few gyri or sulci/ white matter

Global CVI - Involvement of the entire brain

Combined CVI - Combination of the aforementioned types

Images for this section:
Fig. 1: Figure 1. Flow diagrams depicting the normal venous drainage & venous outflow in various pathological states a. Normal Cerebral Venous Drainage b. Venous drainage in venous sinus thrombosis (shaded) c. Venous drainage in venous sinus stenosis d. Venous drainage in arteriovenous malformation e. Venous drainage in spinal dural fistulas. Also, note the classical cord edema involving the dorsal 2/3rds of the cord (star) f. Venous drainage in dural arteriovenous fistula
Fig. 2: Figure 2. Diagram depicting the venous drainage territories a. Axial section at the level of the midbrain shows superior sagittal sinus (blue), sylvian veins (yellow) & vein of Labbe/ transverse sinus (green) territories b. Sagittal section demonstrates the superior sagittal sinus (blue), sylvian veins (yellow) & vein of Labbe/ transverse sinus (green) territories c. Axial section at the level of the basal ganglia & thalamus shows the deep venous (internal cerebral veins & vein of Galen) (orange) territory.
**Fig. 3:** Figure 3. Diagrammatic representation of orbital & parenchymal changes in CVI

a. In carotico-cavernous fistula, there is exophthalmos with orbital congestion. Also, venous reflux into the superficial middle cerebral vein (SMCV) result in CVI b. In children, CVI manifests as hydrodynamic disorder with thickening of the skull vault, enlarged supratentorial ventricular system (star), slit-like 4th ventricle & tonsillar herniation (block arrow) c & d. In "secondary pseudotumor syndrome" there is tortuosity of the optic nerve (curved arrow) with increased fluid in the optic nerve sheath & flattening of the posterior sclera due to papilledema (block arrows).
**Fig. 4:** Figure 4. Proposed Classification of CVI. Focal CVI - Restricted to grey or white matter adjacent to an isolated cortical or medullary vein Local CVI - Restricted to a few gyri or sulci/ white matter Global CVI - Involvement of the entire brain Combined CVI - Combination of the aforementioned types
Imaging findings OR Procedure details

(Figures 5 to 19)

We retrospectively reviewed our interventional neuroradiology database at our institute over the last 15 years from 1997 to 2012 to identify patients with digital subtraction angiographic (DSA) demonstration of CVI. The DSA was performed on Phillips Allura Integris Biplane System. All the patients in the study had undergone cerebral angiogram in atleast Towne's, Lateral and Oblique planes.

All the images of the patients selected for the study were studied in detail to determine any form of CVI. The computed tomography (CT) and magnetic resonance imaging (MRI) findings of these patients, their endovascular management and their follow up was then retrieved from the database. All patients had follow-up of atleast 6 months with atleast one cross-sectional imaging or DSA.

All patients were followed up for a period of atleast 6 months. Our follow-up protocol in all patients undergoing neurointervention involves clinical examination at 1 month, imaging with either ultrasound with Doppler, CT or MRI at 3 month & a follow up with DSA at 6 months.

The imaging features of CVI are myriad. Therefore, we have attempted to describe the findings with a few examples.

In general, since CVI is diagnosed on DSA, the pathognomonic feature of CVI is prolongation of the circulation time. Another important diagnostic feature is the "Pseudo-Phlebitic Pattern" (PPP). The PPP refers to the dilated & tortuous surface veins with contrast stagnation. The other features seen on DSA include meandering veins or veins "hanging-in-space", filling defects in the venous sinuses or cortical vens, venous sinus stenosis, extensive collateralization through alternative paths of venous drainage, venous reflux into veins draining normal brain parenchyma, prominent superior ophthalmic & facial veins, dilation with tortuosity of veins, venous aneurysms, venopathic features like irregular stenosis or dilation of the affected veins etc.

The CT imaging features include hypodensity of the affected brain parenchyma, hemorrhage with "salt-and-pepper"appearance, classical curvilinear hemorrhages, gyral edema with sulcal effacement, parenchymal cacifications in subcortical region, basal ganglia & the brainstem, prominent superior ophthalmic vein, hyperdense sinuses or cortical veins etc
The MRI imaging features include parenchymal edema (T2W - Hyperintensity, T1W - Hypodensity), hemorrhage with "salt-and-pepper" appearance, classical curvilinear hemorrhages, gyral edema with sulcal effacement, parenchymal calcifications in subcortical region, basal ganglia & the brainstem, prominent superior ophthalmic vein, thrombosed sinuses or cortical veins, multiple enlarged collateral venous channels etc

**Images for this section:**

![Images showing various MRI findings](image)

**Fig. 5:** Figure 5. A 39yr-old-man with increasing episodes of left focal seizures. a & b - Venous phase of right ICA angiogram demonstrates filling defect within the collector vein of right parietal developmental venous anomaly with adjacent caput medusae (star). c & d - GRE & T1W images demonstrate adjacent subacute on chronic hemorrhage with thrombosed collector vein (star). e & f - Initial FLAIR & contrast enhanced T1W images show thrombosed collector vein with adjacent hemorrhage (star). g & h - FLAIR & Contrast enhanced T1W images obtained after 3 years show decrease in the size of the hematoma with adjacent thrombosed collector vein of the DVA (star). This is an example of both Congenital Focal CVI (DVAs are due antenatal occlusion of normal venous drainage) & Acquired Focal CVI (collector vein thrombosis with hematoma).
Fig. 6: Figure 6. A 7-month-old-child with sudden onset recurrent right focal seizures. a & b - Left vertebral angiogram reveals a single hole mural type VGAM (vein of Galen malformation). There is extensive venous reflux with flow re-routing and chronic venous ischemia and a classical "pseudo-phlebitic pattern. c, d & e - CT scan reveals dense calcification in the basal ganglia, pons, subcortical calcification & calcification in the wall of the VGAM. f & g - MRI scan reveals heterogenous T1W hyperintensities with perilesional edema due to calcification. h, i & j - TOF MRV reveals extensive cavernous capture of the cerebral venous drainage. This is an example of both Congenital Local CVI (left parietal hematoma) & Congenital Global CVI (parenchymal calcifications).

Fig. 7: Figure 7. A 40yr-old-man with progressive ataxia & imbalance. a, b & c - MRI Brain T2W images show a large partially thrombosed venous aneurysm (notched arrow) with multiple flow voids in the infratentorial fossa. d - On FLAIR
abnormal T2W hyperintensity (arrows) is seen due to venous ischemia. e & f - MRI angiogram reveals enlarged bilateral ECA (occipital) and PCA branches supplying the falcotentorial DAVF (block arrows). g - MR Venogram shows a partially thrombosed torcula (arrowhead) with venous re-routing of the posterior fossa through cerebellar and brainstem veins into the internal cerebral vein. h - CT Brain shows obstructive hydrocephalus. This is an example of Acquired Local CVI (vermian edema).

Fig. 8: Figure 8. A 55yr-old-man with sudden onset headaches & focal seizures. a & b - Right ECA angiograms lateral and AP views show a falcotentorial DAVF draining into an enlarged internal cerebral vein (notched arrow) c & d - CT images show right high parietal bleed (block arrow) with thalamic hypodensity due to focal venous ischemia (arrow) e, f, g & h - T1W, GRE, T2W & FLAIR images show right high parietal bleed (block arrow) with bilateral thalamic hyperintensities (arrow) due to obstruction to the thalamic venous outflow. This is an example of Acquired Local CVI (thalamic edema).
Fig. 9: Figure 9. A 45yr-old-lady with initial complaints of right orbital prominence that worsened to severe headaches & right orbital proptosis after 3 months. a & b - Initial carotid angiograms AP and lateral views demonstrate a very slow flow caroticocavernous fistula on the right with orbital congestion only. c & d - Carotid angiograms after 3 months when patient had sudden worsening of her symptoms. These angiograms reveal venous reflux into the sylvian veins(arrows). e & f - Initial & 3 months later FLAIR images for comparison. The FLAIR images obtained after 3 months show edema in the anterior temporal lobe. g & h - T2W & diffusion images obtained during the flare-up reveals venous edema & congestion with venous ischemic infarct in the anterior temporal lobe. i & j - Initial & 3 months later source images of MR angiograms showing prominent venous reflux into the sylvian veins at 3 months(arrow). This is an example of Acquired Local CVI (temporal edema & infarction).
Fig. 18: Figure 18. A 28yr-old lady with severe papilledema. a & b - Venogram in townes & lateral views showing high grade stenosis in the distal 1/3rd of bilateral transverse sinuses (arrows). c & d - Contrast enhanced MR Venogram demonstrates the high grade stenosis in bilateral transverse sinuses. e & f - T2W images showing flattening of the posterior sclera with increased fluid in the optic nerve sheath. Also, there is empty sella seen. This is an example of Acquired Global CVI (“secondary pseudotumor syndrome”).
**Fig. 17:** Figure 17. A 21yr-old-boy with complaints of "tunnel-vision" in the right eye & blurring of vision in the left eye. Patient was previously diagnosed with extensive sinus thrombosis treated with systemic anticoagulation. a & b- Right ICA angiogram oblique & lateral views show extensive thrombosis of the SSS and the right transverse-sigmoid sinuses with venous re-routing through the cavernous sinuses (star). c & d- T2W images of the right orbit showing both coronal and sagittal "kinking" of the optic nerve sheaths with flattening of the sclera due to "secondary pseudotumor syndrome". Also, there is increased fluid in the optic nerve sheaths. e & f - FLAIR & Post contrast T1W images show non specific T2 hyperintensities in bilateral parietal deep white matter with SSS thrombosis. g & h - Pre and post in situ-thrombolysis venograms show improvement in the caliber of the SSS with decreased venous re-routing through the cavernous sinuses (star) This is an example of Acquired Global CVI ("secondary pseudotumor syndrome").
Fig. 16: Figure 16. A 39yr-old-man diagnosed with falco-tentorial DAVF that was treated with trans-arterial embolization. a & b - Pre procedure T2W axial images at the level of mid pons and basal ganglia. c & d - Post procedure T2W axial images at the level of mid pons and basal ganglia. There is prominent sulcal & sylvian fissure effacement in the pre procedure images when compared to the post procedure images obtained at 3 month follow up. Also, note the marked decrease in the large flow voids (block arrow) adjacent to
the falcotentorium with obstructive hydrocephalus. This is an example of Acquired Global CVI (gyral edema with hydrocephalus).

**Fig. 15:** Figure 15. A 42yr-old-man with sudden onset altered sensorium & chronic headaches. a & b- Left vertebral & ascending pharyngeal angiograms show supply to the falcotentorial DAVF (notched arrow). c, d & e - CT Brain shows multiple calcifications in the basal ganglia & subcortical regions. f, g & h - On the post contrast & CT angiograms multiple serpiginous vessels with enlarged bilateral PCA and ICA branches are seen supplying a falcotentorial DAVF (notched arrow). This is an example of Acquired Global CVI (calcifications).
Fig. 14: Figure 14. A 21-yr-old-boy with complaints of port-wine stain in the left half of face, seizures & right hemiparesis (Sturge -Weber syndrome). a & b - Fluroscopic townes & lateral views of skull demonstrate a thickened calvarium on the left side with tram track calcification in the parieto-occipital lobes. c & d - Venous phase on the left ICA angiogram demonstrates paucity of normal venous structures in the parieto-occipital lobes with venous drainage through deep collateral veins into the SSS. e & f - T2W axial & coronal images show gyral calcifications with left cerebral atrophy & a thickened calvarium on the left side. Also, there is crossed cerebellar diaschisis. g & h - Contrast enhanced T1W axial & coronal images show an enlarged & intensely enhancing left lateral ventricular choroid plexuses compared to the right. This is an example of Congenital Global CVI (antenatal cortical veins fail to develop normally & imaging features are of progressive venous occlusion and chronic venous ischemia).
Fig. 13: Figure 13. A 1yr-old-boy with complaints of left orbital fullness & delayed milestones. a , b & c - Right ICA angiogram reveals a DSM(dural sinus malformation) supplied by bilateral PCAs. There is extensive venous reflux with outflow through the left superior & inferior opthalmic veins (star). c - Venous phase shows high grade bilateral stenosis of the sigmoid - jugular junction(notched arrows). d - CT angiogram shows extensive congestion & tortuosity of the orbital venous system (star). e & f - T1W & T2W images show left sided exophthalmos with dilated SOV & multiple enlarged & tortuous veins in the left temporal fossa (star). g & h - Pre & Post procedural plain CT scans show extensive bilateral high frontal congestion with subcortical calcifications. On the post procedural scan obtained after 1 month, there is increase in subcortical calcification with significant decrease in the venous edema & congestion. Also, note the decrease in the size of the venous pouch with slow flow within. This is an example of Congenital Global CVI (calcifications).
**Fig. 12:** Figure 12. A 2yr-old-girl with developmental delay & regression of milestones. a & b - Left vertebral angiogram reveals a single hole mural type VGAM (vein of Galen malformation) There is extensive venous reflux with flow re-routing and chronic venous ischemia . c, d & e - CT scan reveals dense calcification in the basal ganglia, pons, subcortical calcification & calcification in the wall of the VGAM. f & g - MRI scan reveals heterogenous T1W hyperintensities perilesional edema due to calcification. h, i & j - TOF MRV reveals extensive cavernous capture of the cerebral venous drainage. This is an example of Congenital Global CVI (calcifications).

![Images of various medical scans](image1)

**Fig. 11:** Figure 11. A 35yr-old-lady with complaints of right orbital redness & congestion. a & b- Right ICA angiogram reveals direct CCF with reflux into the right SMCV (superficial middle cerebral vein) (arrow). c & d- CT brain with contrast shows increased number of abnormal vessels in the right perisylvian region. e & f - T2W images show effacement of sulci in the right perisylvian region compared to the left. g & h - CTA & MRA show reflux into the right SMCV (notched arrow). This is an example of Acquired Local CVI (perisylvian gyral edema with sulcal effacement).

![Images of various medical scans](image2)
**Fig. 10:** Figure 10. A 40yr-old-man with severe headaches & generalized tonic-clonic seizures at presentation. 

a & b- Pre and post thrombolysis right ICA angiograms in venous phase reveal antegrade flow through the previously occluded SSS with post thrombolysis decrease in venous ischemia. 

c, d, e & f - FLAIR & T2W images pre and post thrombolysis show dramatic decrease in the bilateral parietal venous edema at 1 month. 

g & h - MR Venogram pre and post thrombolysis showing the dramatic improvement in the flow through SSS at 3 months. This is an example of Acquired Local CVI (parietal lobar edema).
Fig. 19: Figure 19. A 33yr-old-man with lower limb paraparesis. a & b Right L4 lumbar angiogram AP & lateral views show a spinal dural AVF draining into the spinal pial venous plexuses. c, d & e - T2W sagittal & axial images at two successive levels show hyperintensity due to venous ischemia restricted to the posterior 2/3rds of the spinal cord with flow voids on the dorsal aspect of the cord. This is an example of Acquired Global CVI ("venous congestion") in spinal cord.
Conclusion

1. Cross-sectional imaging & DSA play a vital role in the diagnosis & follow-up of patients with cerebral venous ischemia.
2. Though less well characterized, venous ischemia plays an important role in cerebrovascular diseases.
3. The therapeutic goal is resolution of venous ischemia in these patients.

References


Personal Information