CNS injury in acquired metabolic and hydroelectrolytic disorders: a pictorial review of neuroimaging findings

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**Learning objectives**

The main learning objectives of this exhibit are:

1. To review the main imaging patterns of central nervous system (CNS) injury occurring in the setting of acquired metabolic and hydroelectrolytic disorders.
2. To illustrate the imaging spectrum of these pathologies, primarily by magnetic resonance imaging (MRI) but also by computer tomography (CT).

**Background**

As in diseases due to inborn errors of metabolism, CNS may be injured during acquired metabolic and ionic disturbances.

Acquired metabolic disorders can occur in both adults and children, in relation nutritional deficiencies, abnormalities of glucose and electrolyte levels and impaired organ function.

Specific toxins tend to affect specific regions of the CNS selectively, providing recognizable patterns radiologists should be familiar with.

We collect a series of cases from our Neuroradiology imaging archive, illustrated by MR and CT imaging. A small introduction to each pathology and classical clinical and imaging findings is provided.

**Findings and procedure details**

**1 - Hypoglycemia**

- Neonatal hypoglycemia (<46 mg/dL) occurs in 5%-15% of normal term neonates and can cause visual impairment, epilepsy, and cognitive deficits.
- Most common metabolic imbalance of the newborn.
- In the adults is usually related to overmedication with oral antidiabetic drugs.

- Imaging:
  
  • **Neonatal Form**
  
  - Typical location of lesions: Bilateral occipital/parietal lobes (cortex and white matter)
  
  - Also documented by imaging and/or pathology
  
  - Frontal and temporal lobes, basal ganglia, thalami, brain stem, posterior limb of the internal capsule can be affected.
  
  - **CT:** Hypodense, it may have hemorrhage in acute phase or calcifications in chronic phase
  
  - **MRI:** Acute/subacute phase: T1 and T2 loss of cortical signal or hypersignal, T2 white matter hypersignal, it may have hemorrhage; DWI hypersignal with ADC reduction in acute phase (Fig. 1 on page 7).
  
  - Chronic phase: encephalomalacia, atrophy, generally with T2 hypersignal (Fig. 2 on page 7).

  • **Adult/Non-Neonatal Form**
  
  - Typical location of lesions: Bilateral cerebral cortex (mainly parietal, occipital, temporal), hippocampi, amygdalae
  
  - Basal ganglia (globus pallidus and/or striatum), subcortical white matter
  
  - **CT:** Diffuse edema, hypodense
  
  - **MRI:** Acute/subacute phase: laminar necrosis (T1 hypo or hypersignal, T2 hypersignal, DWI hypersignal with ADC reduction, may enhance with contrast); reversible ADC reduction
  
  - Chronic phase: atrophy, with or without signal modification; total resolution

2 - Nonketonic hyperglycemia

- Nonketotic hyperglycemia affects patients with poorly controlled diabetes mellitus

- Classically present with clinical finding of Hemichorea-HemiBallism.

- Imaging
  
  • Characteristic appearance of unilateral or asymmetric lesions of the basal ganglia (BG) contralateral to the side of patient's symptoms (Fig. 3 on page 8).
  
  • **Non-contrast CT:** hyperdense putamen and/or caudate nucleus contralateral to the side of symptoms.
  
  • **MRI:** high signal intensity BG lesions on T1-weighted.
  
  • Putamen is almost always involved with variable additional associated BG lesions.
• T2-weighted findings are much more variable.

3 - Kernicterus

- Kernicterus is a neurological manifestation of hyperbilirubinemia in the newborn, with premature neonates being more susceptible.

- Results from cerebral deposition of unconjugated bilirubin -

- Presents typically in the newborn (2-5 days-old) with jaundice, lethargy, hypotonia and high-pitched cry.

- Serum bilirubin levels are elevated (>20mg/dL)

**Imaging:** MRI

- May be normal.
- Typical location of lesions: globus pallidus (GP) (most common), subthalamic nuclei and hippocampus; less frequently: thalamus, striatum, substantia nigra, cerebellar nuclei and cranial nerves. Cerebral cortex and white matter are classically spared.
- Acute phase: increased SI on T1WI (Fig. 4 on page 9).
- Subacute/chronic phase: T2 hypersignal, atrophy.
- Lesions are symmetrical.

Treatment: Phototherapy and exchange transfusion in severe cases.

4 - Disturbances of calcium/phosphorus metabolism

- There can be acquired disturbances of phosphorus and calcium metabolism.

- Usually associated to hormonal imbalances (primarily PTH).

- Imaging:

  - Typical location of lesions
  - Bilateral basal ganglia (globus pallidus and/or putamen and/or caudate), thalami, dentate nuclei, subcortical/central white matter, duramater.
  - **CT:** Calcifications 
  - **MR:** T1 hypersignal, T2 variable (mostly hyposignal), T2* marked hyposignal ("blooming")
5 - Acquired Hepatocerebral Degeneration

- Chronic hepatic encephalopathy (HE)
- Spectrum of neuropsychiatric abnormalities occurring in patients with liver dysfunction.
- Most cases associated with cirrhosis, but an also be seen in patients with acute liver failure and, rarely, with portal-systemic bypass
- Results from accumulation of Manganese and other paramagnetic substances

- Imaging:
  - Typical location of lesions: Bilateral GP extending to anterior midbrain (mostly the internal portion of cerebral peduncle /substantia nigra)
  - May affect: Adenohypophysis (Fig. 7 on page 11), striatum, subthalamic nuclei, hypothalamus, tegmentum, cortex and white matter
  - MRI: T1 hypersignal, T2 normal (or hypersignal); cortex and white matter lesions have more frequently T2 hypersignal (Fig. 6 on page 11)
  - It may disappear with normalization of hepatic function

6 - Wernicke encephalopathy (alcoholic/nonalcoholic)

- WE is an acute neurologic disorder resulting from thiamine (vitamin B1) deficiency.
- When untreated, severe neurologic deficits like Korsakoff psychosis and even death may ensue.
- Despite being preventable and treatable it remains underdiagnosed

- Imaging:
  - Typical location of lesions: Bilateral periaqueductal gray matter and periventricular region of the third ventricle (mamillary bodies, medial thalami, tectal plate).
  - Pontine and medulla tegmentum, cranial nerves nuclei (VI, VII, VIII, XII), red nuclei, cerebellar hemispheres and vermis, dentate nuclei, hypothalamus, putamen, caudate, cortico-spinal tracts, cerebral cortex (mostly central and peri-central cortex), splenium, fornix.
  - CT: Hypodense (mostly in medial thalami).
  - MRI: Acute phase: T1 hyposignal or normal, T2 hypersignal, DWI hypersignal with ADC reduction, it may enhance with contrast (mainly in
mamillary bodies, medial thalami, tectal plate, periaqueductal) (Fig. 8 on page 12, Fig. 9 on page 13).

- Chronic phase: atrophy, mainly in mamillary bodies; normalizes with correction of vitamin deficit.

7 - Osmotic demyelination (pontine/extrapontine)

- Acquired condition that results in an osmotic insult and demyelination in the setting of rapid correction of hyponatremia.

- Although the pontine center represents the most common site of involvement, lesions do occur outside of the pons and are termed Extrapontine osmotic demyelination.

- More common in patients with prolonged hyponatremia, history of chronic alcohol abuse and malnutrition.

- Imaging:
  - **CT:** low density lesions in the pons or other affected regions, and occasionally shows enhancement
  - **MRI:** Acute phase: T1 hyposignal or normal, T2 hypersignal, DWI hypersignal with ADC reduction, it may enhance with contrast
  - Chronic phase: normalizes with correction of the basal condition

a) Pontine form

Typical location of lesions: Central pons, with a triangular or round/oval shape; spares the peripheral fibers and cortico-spinal tracts (Fig. 10 on page 15).

b) Extrapontine form

Typical location of lesions: Bilateral basal ganglia (more common), cerebral or cerebellar white matter, cortex, thalami, hippocampus, lateral geniculate body (Fig. 11 on page 14).

8 - Subacute combined degeneration

- Vitamin B12 deficiency is a systemic disease that often affects the nervous system.

- One of the most prevalent manifestations is subacute combined degeneration (SCD) of the spinal cord.
- It’s a well defined disorder with dysaesthesia, disturbance of position sense, and spastic paraparesis or tetraparesis.

- Corresponding neuropathological findings are a diffuse, multi-focal pattern of axonal loss and demyelination most severe in the cervical and thoracic spinal cord.

- Predominantly affects the posterior columns followed by the anterolateral and anterior tracts.

- Imaging:
  - Typical location of lesions: Posterior columns of spinal cord in a "inverted V" shape; mainly in lower cervical and upper thoracic segments.
  - Lateral and sometimes anterior columns; all spinal cord; brain stem tegmentum.
  - **MRI**: Acute phase: T2 hypersignal, T1 normal or slightly hypointense, it may enhance (Fig. 12 on page 16).
  - Chronic phase: normalizes with correction of vitamin deficit.

Images for this section:

![Fig. 1:](image1)

**Fig. 1:** Newborn with hypoglycemia. ADC map (A) and DWI (B) show restricted diffusion in the occipital cortex. On T2WI (C) slight effacement of the occipital sulci can be seen.
Fig. 2: Sequelae of hypoglycemia. On A (newborn shown on Fig. 1) only a very discreet hyperintensity of the occipital white matter can be seen. He had no neurological deficits. B shows a different child with areas of encephalomalacia after hypoglycemia.
Fig. 3: (A) Axial non-contrast CT demonstrates interval hyperdensity within the left caudate and lentiform nucleus (arrow) which respects the neuroanatomic boundaries of the basal ganglia without associated edema. There is hyperintensity of the same areas on T1WI (B, D), with normal SI on T2WI (C).
Fig. 4: Magnetic Resonance Imaging obtained at 8 days of age shows bilateral high signal intensity (SI) on the globus pallidus (GP) and subthalamic nuclei on axial T1WI (A, B, arrow head) and of the GP on T2WI (C, white arrow). Hyperintensity of hippocampal cortex (D, black arrow) can also be seen in T2WI. (E) T2WI of 8-day-old newborn with normal hippocampal cortex SI.
**Fig. 5:** CT scan of a patient with hypoparathyroidism. There are calcifications of the basal ganglia and subcortical white matter. There is no edema or mass effect.

![CT scan images](image)

**Fig. 6:** Typical findings in chronic hepatic encephalopathy of T1 high signal intensity of the GP extending inferiorly to the mesencephalon.
**Fig. 7:** T1WI. High signal intensity of the adenohypophysis in a patient with hepatic encephalopathy.
Fig. 8: 34-year-old man with a history of food refusal presented with altered consciousness. (A) axial T2W images show high signal-intensity in the medial thalami (orange arrows) and periaqueductal area (black arrow). (C, D) DWI - Note signal intensity alterations in the mammillary bodies (arrow head), and thalami (white arrows).
Fig. 9: Alcoholic patient with Wernicke encephalopathy. T2W FLAIR images (A-E) show high signal intensity of the posterior aspects of the brainstem (arrows). DWI image (F) shows restricted diffusion on the tectal region.
**Fig. 11:** T2-weighted image shows diffuse involvement of basal ganglia (arrows) characteristic of extrapontine myelinolysis.
Fig. 10: 56 year-old alcoholic man after rapidly corrected hyponatremia. (A) CT image shows hypodensity of the central pons (arrow). (B) T2-weighted image shows signal prolongation in pons (arrow) typical of central pontine myelinolysis. Diffusion-weighted image (DWI) (C) and apparent diffusion coefficient map (D) show restricted diffusion (arrows).
Fig. 12: T2WI showed hyperintense lesions within the dorsal columns. T1 weighted images (not shown) were normal and no enhancement was seen after intravenous administration of gadolinium.
Conclusion

Overall, acquired metabolic and hydroelectrolytic disorders preferentially originate lesion in the deep gray matter, with less frequent involvement of white matter and cortex; they are typically bilateral and symmetrical, with the exception of nonketotic hyperglycemia, which is usually unilateral.

Although imaging patterns are not totally specific, imaging studies and mainly MRI can show suggestive lesions.

Most typical location of lesions and atypical or less frequent locations should be recognized.

Recognizing these patterns might be life-saving, as in the case of Wernicke encephalopathy or prompt adequate and timely treatment for other conditions.

Personal information

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