Learning Objectives

Group
deep grey matter signal changes based on their signal characteristics and location

Recognize
characteristic patterns and signs that can help narrowing the differential

Review
different underlying etiologies and typical clinical presentations

Describe the myofascial cone of the orbit and how it applies to the orbital spaces and imaging.

Compare and contrast how MRI and CT are used to evaluate orbital pathology.

Identify common and important orbital pathologies based on their imaging appearance and location.

Group deep grey matter signal changes based on their signal characteristics and location.

Recognize characteristic patterns and signs that can help narrowing the differential.

Review different underlying etiologies and typical clinical presentations.
Background

The deep grey matter (DGM) is rich in mitochondria, neurotransmitters and vascular supply. Increased glucose and oxygen metabolism makes it vulnerable to metabolic, ischemic and toxic abnormalities.

**Conditions affecting deep grey matter**
- toxic poisoning
- metabolic
- degenerative
- vascular
- inflammation/infection
- malignancy

There is variation and overlap in radiological and clinical features, therefore, imaging, labs and clinical data should be correlated to reach the correct diagnosis.
MR signal changes in the deep grey matter
**Chronic Hepatic Encephalopathy**

- **Manganese** deposition due to hepatic failure.
- **T1 bright** bilateral globi pallidi.
- Bilateral bright T1 globi pallidi can also be present due to copper accumulation (Wilson disease) and total parenteral nutrition.
- **Symptoms**: cognitive impairment, movement disorder.

**Diabetic Striatopathy**

- **T1 bright** unilateral striatum.
- Hyperglycemia, hyperosmotic state.
- **Symptoms**: involuntary movements (hemiballismus, hemicoreia)

Cases:
- Case: Frank Gaillard rID: 46014
- Case: Ayaz Hidayatov rID: 56241
High T2 signal in bilateral basal ganglia. Cortical and subcortical white matter might be affected.

Complex metabolic changes due to renal failure

Symptoms: movement disorder, altered mental status, seizures

Lentiform fork sign
Hyperintense rim around putamen and globus pallidus thought to be resulting from metabolic acidosis.

Case: Coenraad Hattingh rID: 37856
Happens by drinking windshield wiper fluid or moonshine leading to metabolic acidosis.

Relative sparing of globus pallidus is typical. Optic nerve, subcortical white matter can be affected. Diffusion restriction can occur in the acute phase.

**Symptoms:** GI and visual symptoms, rapidly progressing to coma
Copper deposition

Liver cirrhosis with CNS manifestation affecting the basal ganglia and midbrain.

Globus pallidus are T1 bright if hepatic failure dominates or T1 dark if edema is present.

**Double panda sign**

Face of a giant panda Miniature panda

Caused by Wilson disease

Case: Frank Gaillard rID: 4438

Globi pallidi are T1 bright if hepatic failure dominates or T1 dark if edema is present.

**Symptoms:** Parkinsonism, dystonia, dysarthria, ataxia. Kayser-Fleischer rings might be present

**Case:** Paresh K Desai rID: 12107

**Putamen**
- Uremic encephalopathy
- Methanol poisoning
- Wilson disease

**Globus pallidus**
- Carbon monoxide poisoning

**Thalami**
- Wernicke's encephalopathy

**Dentate nuclei**
- Metronidazole toxicity
Carbon monoxide causes anoxia due to its higher affinity to hemoglobin than oxygen.

Poisoning occurs due to idling vehicles, wood burning fireplaces, unvented space heaters.

**Bilateral** symmetric necrosis of *globi pallidi* appearing as high T2 signal. White matter involvement can occur. **Diffusion restriction** is seen in the **acute phase**.

**Symptoms:** Headache, dizziness, seizures, coma.
Thiamin (B1) deficiency

Due to malnutrition, alcoholism, hyperemesis, bariatric surgery, etc.

Symptoms: Triad of acute confusion, ataxia, opthalmoplegia. **Korsakoff psychosis** is its chronic form with confabulation and memory loss.
High T2 signal in dentate nuclei, splenium of corpus callosum, tegmentum, dorsal brainstem.

Symptoms: Cerebellar dysfunction, altered mental status, seizures

Antibiotic with anaerobic and protozoal coverage. Prolonged use can lead to neurotoxicity.
An autosomal recessive neurodegenerative disease presenting during the first two decades of life.

Iron deposition appears as signal drop on T2, T2* and SWI. Bilateral globi pallidi and pars reticulata of the substantia nigra are affected.

Symptoms: Spasticity, progressive dementia, extrapyramidal signs. Progressive with death in the second or third decade.
A neurodegenerative synucleinopathy previously known as striatonigral degeneration with predominantly parkinsonian features.

Low T2 and T2* signal in putamen. Reduced putaminal volume.

**Putaminal rim sign**
Linear high T2 rim surrounding the putamen only seen at 1.5T

**Symptoms:** parkinsonism, mild cerebellar and pyramidal signs starting between 40 and 60 years of age.
Calcium deposits in the capillary walls in the globus pallidus, putamen, caudate, thalamus, dentate nucleus, corona radiata and subcortical white matter causing low T2/T2* signal and hyperdensity on CT.

Small amount of age-related basal ganglia calcification is considered normal.

**Fahr disease**: idiopathic, primary familial brain calcification, presents at 40-60 years.

**Fahr syndrome**: secondary due to various causes of hypercalcaemia (e.g. hypoparathyroidism, infections, etc.)

**Symptoms**: often asymptomatic. Progressive psychosis, cognitive impairment, dementia, gait disturbance, movement disorders.
Diffusion restriction in the basal ganglia can be a result of various conditions including ischaemia, metabolic changes such as hypoglycemia, infection or hypercellular tumors such as primary CNS lymphoma.

The differential for bilateral thalamic diffusion restriction is worth to be aware of.

**Bilateral thalamic diffusion restriction differential**

**Ischemia**
- Arterial - artery of Percheron infarct, tip of basilar infarct
- Venous - straight sinus thrombosis, deep cerebral vein thrombosis

**Metabolic**
- Wernicke's encephalopathy

**Infection**
- Creutzfeldt-Jacob disease
- Flavivirus encephalitides
**Artery of Percheron Infarct**

The thalami are supplied by perforators arising from the PCA. Artery of Percheron is a rare variant when a solitary arterial trunk arising from PCA supplies bilateral paramedial thalami and the midbrain. Occlusion results in bilateral symmetric thalamic infarcts.

**Symptoms:** Triad of **vertical gaze palsy**, altered mental status, memory impairment. Oculomotor disturbances, movement disorder, cerebellar ataxia and movement disorder in case of midbrain involvement.
**CREUTZFELDT-JACOB DISEASE**

Prion disease causing spongiform encephalopathy.

There are four different types including **sporadic** (85-90% of all cases), **variant** (bovine-to-human), **familial** and **iatrogenic** (related to neurosurgery or cadaveric pituitary hormones, etc.).

**Diffusion restriction** in the basal ganglia and cerebral cortex is characteristic.

**Symptoms:** Progressive cognitive decline and death.

- **Pulvinar sign**
  - Symmetrical hyperintensity of the pulvinar thalamic nuclei only.

- **Hockey stick sign**
  - Symmetric hyperintensity of pulvinar nuclei and dorsomedial thalami.

- **Cortical ribboning**
  - Diffusion restriction in cerebral cortex.

---

**Bilateral thalamus**

- Percheron infarct
- Creutzfeldt-Jacob
- Flavivirus encephalitis

**Other conditions**

- Hypoxic ischemic encephalopathy
- Hypoglycaemia

---

Case: Paloma Martínez Sebastiá

rID: 86996

Case: Andrea Horváth

rID: 169236

Case: Chris O'Donnell

rID: 16320
Many different flaviviruses can cause encephalitis including Japanese encephalitis virus, West Nile or dengue fever viruses.

Symmetric basal ganglia and/or thalamic diffusion restriction and T2 hyperintensity is characteristic.

**Symptoms:** Prodrome including fever, rash, myalgia, headache. Neurological symptoms include tremor, dystonia, dyskinesia, dysarthria, seizures.
**HYPOXIC ISCHEMIC ENCEPHALOPATHY**

**HYPOGLYCEMIC ENCEPHALOPATHY**

Prolonged global hypoxia due to respiratory or circulatory failure. Hypoglycemia can result from hypoglycemic medication overdose or insulinoma.

**Diffuse** cortical and deep grey matter involvement. Involvement of hippocampi, internal capsules (posterior limb) and parieto-occipital cortex is also characteristic in hypoglycemia.

Cytotoxic edema secondary to cell death results in **diffusion restriction** and eventually in high T2/FLAIR signal. Diffuse cerebral hypoattenuation, loss of grey-white differentiation and swelling is seen on CT.

**Symptoms:** Altered mental status

**Case:** Andrew Dixon  rID: 38998
Various other conditions can cause signal changes in the basal ganglia. Evaluating them in the appropriate clinical context is critical. Here are few examples.

**AGE-RELATED IRON OR CALCIUM DEPOSITION**
Dilated Virchow-Robin spaces.

**ÉTAT CRIBLÉ**

**MALIGNANCY - GLIOMA**

**TOXOPLASMOsis**

---

Hegde et al., Radiographics. 2011 Jan-Feb;31(1):5-30.

Case: Roberto Schubert
rID: 1381

Case: Bianca Larissa Pereira
rID: 66119

Case: Frank Gaillard
rID: 30082
SUMMARY

- High T1: Chronic Hepatic Encephalopathy, Diabetic Striatopathy
- High T2: Uremic Encephalopathy, Methanol, Wilson Disease, Carbon Monoxide, Wernicke's, Metronidazole
- Low T2/T2*: PKAN, MSA-P, Fahr's
- Diffusion Restriction: Artery of Percheron Infarct, Creutzfeldt-Jacob, Flavivirus Encephalitis, Hypoxia Hypoglycemia
A high variety of conditions can result in signal changes in the basal ganglia with the majority of them being toxic or metabolic. This is due to the high mitochondrial content of grey matter as a consequence of high metabolic demand of neuronal activity.

It is worth to be aware of specific patterns to narrow the differential. Categorizing these conditions based on signal changes and location can be a useful tool.

Interpreting imaging in combination with the clinical and laboratory findings can be helpful as the presentation or radiological picture is often non-specific.
References

- Sun, Shi and Mandell: Core radiology
- Prometheus Lionhart: Crack the core exam
- radiopaedia.org
- The “double panda sign” in Wilson’s disease