Agenda

➢ Our patient Clinical history

➢ Anatomy of the Basal Ganglia

➢ Pathophysiology

➢ Differential diagnosis

➢ Imaging of diseases causing BGC

➢ Summary
Our Patient Clinical history

A Female patient, HTN, was found down in her bed unresponsive.

History notable for 4L coffee ground emesis at OSH and a fever as well as elevated CK and transaminases. Neurologically she is grossly nonfocal but does have significant cognitive slowing and difficulty with more complex commands.

The patient has had a prodrome of personality changes, specifically apathy and seeming depression four month ago.
Findings:

Bilateral high attenuation areas in the Basal Ganglia representing calcified areas.

Other findings:
Choroid plexus calcification
Findings:

Low attenuation areas in both Globus pallidus

Other Findings:

Choroid plexus calcification

Source: “PACS, BIDMC”
Findings:

High intensity of Globus Pallidus
Anatomy:

Basal Ganglia:

- Caudate nucleus
- Putamen
- Globus Pallidus

Illustration of the basal ganglia structures
Anatomy:

Basal Ganglia nuclei Grouped together:

Striatum: Caudate + Putamen

Lentiform: Globus pallidus + Putamen

Corpus Striatum: Lentiform + Caudate
Pathophysicsiology:

- Calcium interaction with fatty acids
- Rupture of Blood Brain Barrier
- Iron may play a role as it catalyzes reactive oxygen radicals
- Elevated intracellular Calcium
Differential Diagnosis:

**Idiopathic:**
- Ageing
- Fahr disease

**Toxic:**
- Carbon monoxide
- Lead
- Mineralizing microangiopathy
- Anticonvulsant therapy

**Infectious:**
- CNS Tuberculosis
- AIDS
- Neurocysticercosis
Differential Diagnosis:

**Metabolic:**
- Hypoparathyroidism
- Pseudohypoparathyroidism

**Inherited:**
- Mitochondrial disease as MELAS
Differential Diagnosis:

Idiopathic:
  • Ageing
  • Idiopathic Fahr disease
Ageing:

- Usually idiopathic, with an incidence rate of 1%
- Age of presentation seems to regulate the type of symptoms expressed by affected patients.
- Incidence of neuropsychiatric findings is most dependent on amount of mineralization.
- 50% of patients with extensive brain mineralization exhibited mental disorders.
Differential Diagnosis:

Idiopathic:
• Ageing
• Idiopathic Fahr disease
Fahr Disease:

- Idiopathic Basal Ganglia Calcification

- Presents in 4\textsuperscript{th} and 5\textsuperscript{th} decade.

Symmetric, bilateral involvement of:

- Globus pallidus
- Caudate
- Lentiform nucleus
- Thalamus
- Dentate nucleus

- MRI T1 show high signal.

- PET scan may show decrease FDG uptake.
Fahr Disease:

Findings:
Bilateral Dentate nucleus Calcification

Other findings:
Pineal gland calcification

Companion patient 1: Axial CT non contrast:

Courtesy of Dr Rafael Rojas
Fahr Disease:

Findings:

Bilateral Basal ganglia and Dentate nucleus calcification
Fahr Disease:

Companion patient 1: Axial CT non contrast:

Findings:

Bilateral Corpus Striatum & subcortical calcification
Fahr Disease:

Companion patient 1: MRI T1 and T2:

[Images of MRI scans showing abnormalities]
Fahr Disease:

Companion patient 1: MRI Flair and T1:

Courtesy of Dr Rafael Rojas
Differential Diagnosis:

Toxic:
- Carbon monoxide
- Lead
- Mineralizing microangiopathy
- Anticonvulsant therapy
CO poisoning:

- Carbon monoxide binds to hemoglobin approximately **200 times** more tightly than oxygen.

- The neurotoxicity of CO could be acute or chronic.

- Globus pallidus is the most affected area.

- Classically seen as low attenuation of globus pallidus on CT, low signal on MRI T1 weighted imaging and high signal on T2/FLAIR.
CO poisoning:

Findings:

Bilateral Globus pallidus low attenuation.
CO poisoning:

Findings:

Bilateral globus pallidus oval shaped areas of altered signals eliciting low T1

Source: http://radiopaedia.org/cases/carbon-monoxide-poisoning-1, Dr Muhammed Essam
Differential Diagnosis:

Toxic:
- Carbon monoxide
- Lead
- Mineralizing microangiopathy
- Anticonvulsant therapy
Lead toxicity:

Findings:

Hyperintense signal alterations of the basal ganglia

Differential Diagnosis:

Toxic:
- Carbon monoxide
- Lead
- Mineralizing microangiopathy
- Anticonvulsant therapy
Mineralizing Angiopathy:

• Usually presents in children receiving Chemo or radiotherapy, but other causes are possible as trauma.

Areas mostly affected include:
• Corticomedullary junction
• Lentiform nucleus
• Dentate nucleus of cerebellum
Mineralizing Angiopathy:

Findings:

Hyperdense areas noted in the basal ganglia and sub-cortical white matter
Differential Diagnosis:

**Infectious:**
- CNS Tuberculosis
- AIDS
- Neurocysticercosis
CNS Tuberculosis:

- Tuberculosis is caused by mycobacterium tuberculosis.
- The disease begins with the development of small tuberculous foci (Rich foci) in the brain, spinal cord, or meninges.
- **CT** non-contrast scans may be normal
- **MRI** T1 gadolinium enhanced shows hyperintensity
CNS Tuberculosis:

Findings:

Multiple lesions involving the cerebral hemisphere including the basal ganglia.
Differential Diagnosis:

**Infectious:**
- CNS Tuberculosis
- AIDS
- Neurocysticercosis
AIDS:

• AIDS is caused by infection of HIV, which affects CD4+ cells.
• AIDS affects the basal ganglia early in the disease as evidenced by slow cognition and motor reaction times even in asymptomatic HIV positive patients.
AIDS:

Findings:

Bilateral basal ganglia calcification.
Differential Diagnosis:

**Infectious:**
- CNS Tuberculosis
- AIDS
- Neurocysticercosis
Neurocysticercosis:

- Caused by ingestion of Tenia solium eggs.

- Larval cysts commonly found in the central nervous system but they can also be found in the eye, muscle or other tissues.

- Findings are variable on CT, but most prominent during the calcified stage.

- MRI is the modality of choice to view Neurocysticercosis
Neurocysticercosis:

Findings:

Hyper intense lesion affecting the right putamen and left caudate.

Source: Clinical Neurology and Neurosurgery 104 (2002) 57–60
Differential Diagnosis:

Metabolic:
- Hypoparathyroidism
- Pseudohypoparathyroidism
Hypoparathyroidism

- Decreased PTH levels causing ↓ Ca & ↑ P.
- Increase P levels causes Ca to deposit in the brain tissue.
- Bilateral, Symmetrical
- Affects grey-white junction, Cerebellum
- Non Contrast CT has highest sensitivity and specificity
- MRI not useful as signal intensity of calcified lesion varies widely.
Hypoparathyroidism

Findings:

Bilateral Lentiform high attenuation.

Other Findings:

Bilateral thalamus, and multiple subcortical lesions. Choroid plexus calcifications.

Differential Diagnosis:

Metabolic:
• Hypoparathyroidism
• Pseudohypoparathyroidism
Pseudohypoparathyroidism

- Pseudohypoparathyroidism is a condition associated with resistance to parathyroid hormone.
- Subtypes:
  type I: abnormal cAMP response to PTH stimulation
  - type Ia: has characteristic phenotypical features
  - type Ib: lacks phenotypical features
  type II: normal cAMP response to PTH stimulation
- Affects deep white matter and basal ganglia
Pseudohypoparathyroidism:

Findings:

Extensive basal ganglia and cerebral calcification
Differential Diagnosis:

Inherited:

• MELAS
**MELAS:**

- Mitochondrial encephalo- myopathy, lactic acidemia, and stroke like symptoms.

- Mitochondrial disease of maternal inheritance.

- Symmetric basal ganglia calcification

- Focal cerebral lesions not confined to the vascular territories in a young patient.

- Muscle biopsy may show ragged fibers.
MELAS:

Findings:

Bilateral Lentiform nucleus hyperintensity

Summary

• The best modality is CT non Contrast.

• Incidental findings are common with age.

• The most common area affected is Globus pallidus.

• Most likely mechanism is disruption of Blood Brain Barrier.

• The most common cause is Fahr disease and metabolic disorders.
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References:

- **M Mej doub i**, J Neurol Neurosurg Psychiatry. Dec 2006; 77(12): 1328
- Clinical Motor and Cognitive Neurobehavioral Relationships in the Basal Ganglia, By Gerry Leisman, Robert Melillo and Frederick R. Carrick
References:

- www.radiopaedia.org/cases/carbon-monoxide-poisoning-1, Dr Muhammed Essam
- www.radiopaedia.org/cases/carbon-monoxide-poisoning, Dr Ruslan Esedov
- www.radiopaedia.org/cases/mineralising-microangiopathy, Dr Ayush Goel