Imaging of basal ganglia abnormalities

Poster No.: C-1586
Congress: ECR 2014
Type: Educational Exhibit
Authors: M. Hachicha¹, H. Abid², M. Ben Lassoued², B. Souissi², N. Toumi², A. Maalej², S. Haddar², K. Ben Mahfoudh³; ¹Kairouan/TN, ²Sfax/TN, ³3000/TN
Keywords: Neuroradiology brain, MR, Diagnostic procedure, Metabolic disorders
DOI: 10.1594/ecr2014/C-1586

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR's endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

- The deep grey matter structures of the basal ganglia comprise the caudate nucleus, putamen and globus pallidus. They form the key components of the extrapyramidal motor system, and receive projections from almost every region of the cerebral cortex, playing a vital role in integrating movement.

- They have high energy [adenosine triphosphate (ATP) produced by oxidative phosphorylation within the mitochondria] requirements, increased blood flow and are rich in neurotransmitters and trace metals such as iron, copper and manganese. Hence, they are vulnerable to any systemic disease or generalised process.

- Abnormalities of the basal ganglia and thalamus may be detected at neuroimaging in a wide variety of pathologic conditions classified as systemic or focal, some with acute onset and others with slowly progressive manifestations.

- Magnetic resonance (MR) imaging is the modality of choice for evaluating the basal ganglia. The neuroradiologist can thus play an important role in contributing imaging features to the overall clinical, biochemical and genetic picture that makes up an accurate picture of patients with systemic and metabolic disease.

- The objective of our study was to establish the value of MR with new techniques (diffusion-weighted imaging and MR spectroscopy) in the study of abnormalities of the basal ganglia.

- In this work we describe bilateral basal ganglia lesions from different causes using illustrative examples to highlight neuroimaging and non-imaging features that may be helpful in distinguishing some of them.
Background

I-MRI Appearance of Basal Ganglia Lesions:

-the caudate nucleus and putamen are isointense relative to the cortical gray matter with all pulse sequences and do not enhance after contrast material injection.

-The globus pallidus is typically slightly hypointense relative to the putamen, a normal feature that is attributable to progressive iron deposition as one ages.

-In paediatric patients, immature myelination may result in indistinct intensity differences, but with normal maturation and progressive (physiological) deposition of iron, there is progressive shortening of the T2 signal intensity, resulting in the decrease of signal intensity on T2-weighted images. This process occurs first in the globus pallidus followed by the putamen.

- Most pathology in the basal ganglia with high signal lesions on T2-weighted images have a corresponding decreased signal on T1-weighted images, but there are other pathology with hyperintense signal on T1-weighted images, notably Wilson’s disease, hepatic failure and in hyperglycaemia. Diffusion-weighted (DW) MR imaging and MR spectroscopy (MRS) may be helpful in acute metabolic disease.

II-Illustrative Examples:

Most examples of bilateral basal ganglia lesions in this work are taken from our department of radiology (during 3 years (2009-2011)), and are not meant to be exhaustive.

II.1.Metabolic and toxic causes:

II.1.1.Fahr Disease(Figure1):

- Fahr Disease is a rare neurodegenerative disease that is characterized by the bilaterally symmetric deposition of calcium (and other minerals) in the basal ganglia, thalamus, dentatenuclei, and centrum semiovale in the absence of hypoparathyroidism.

- Clinical signs:

  * non specific gradual onset symptoms such as headache, vertigo, movement disorders, syncope, and seizures.

  * Other neurologic deficits include paresis, spasticity, gait disturbance, speech disorders, coma, dementia, Parkinsonism, chorea, tremors, dystonia, myoclonia, and orthostatic hypotension.
- Imaging: * bilaterally symmetric dense calcifications in the basal ganglia, dentate nuclei, thalamus, and subcortical white matter of the cerebrum

* Brain CT scan, which easily detects calcium, is the preferred method to localize and assess the extent of cerebral calcifications. On MRI, calcified areas in the basal ganglia give a low-intensity signal on T2-weighted images and a low- or high-intensity signal on T1-weighted planes.

II.1.2. Wilson Disease(Figure2):

- Wilson disease also called hepatolenticular degeneration, is caused by the accumulation of copper resulting from a deficiency of ceruloplasmin, its serum transport protein.

- This disease, affects the liver, brain, and other tissues.

- Clinical signs: dysarthria, dystonia, tremors, ataxia, Parkinsonian symptoms, and psychiatric problems.

- Kayser-Fleisher rings in the cornea are characteristically associated with Wilson disease.

- MRI: * areas of T2 prolongation in the putamen (a common finding), globus pallidus, caudate nuclei, and thalamus.

* Thalamic involvement is typically confined to the ventrolateral aspect. The cortical and subcortical regions, mesencephalon, pons, vermis, and dentate nuclei may also be involved.

* Diffusion restriction is often seen in the early stages of the disease

II.1.3. Wernicke Encephalopathy(Figure3):

- Typically results from a vitamin B1 (thiamine) deficiency, secondary to a malnourished state caused by (for example) chronic alcoholism, gastrointestinal or hematologic neoplasms, chronic dialysis, bowel obstruction, hyperemesis gravidarum, or prolonged parenteral therapy without vitamin supplementation.

- Clinical triad: altered consciousness, ocular dysfunction, and ataxia

- Wernicke encephalopathy represents a medical emergency, and treatment consists of intravenous replacement of thiamine

- MRI: symmetric T2 prolongation in the medial thalamus, periaqueductal area, mamillary bodies, and tectal plate. Petechial hemorrhage, diffusion restriction, and contrast enhancement of the affected areas may be noted.

- ADC: normal (early stage), low ADC (late stage) in relation to irreversible damage.
II.1.4. Acute Liver Disease (Figure 4 and 5):

- Characteristic MR imaging signs of acute brain damage may be present in patients with acute hyperammonemia, including cirrhotic patients with acute hepatic decompensation and patients with ornithine transcarbamylase deficiency (ie, inborn errors of metabolism such as citrullinemia, which result in accumulation of ammonia in the brain).

- MRI: *bilateral symmetric swelling, T2 prolongation, and restricted diffusion in the basal ganglia, insular cortex, and cingulate gyrus.

* MR spectroscopic detection of the combined toxic metabolite glutamate-glutamine at short echo times

II.1.5. Toxic Cause: Carbon Monoxide Poisoning (Figure 6):

- Carbon monoxide (CO) poisoning is the most common cause of poisoning morbidity and mortality.

- Clinical signs: cognitive impairment, coma and death.

- Sequelae: in survivors include cognitive damage, dementia and Parkinsonian features.

- Pathological effects of carboxyhaemoglobin on the brain include necrosis, acute demyelination and chronic atrophy.

- The globus pallidus is the most common site of bilateral symmetrical abnormality.

- Whole basal ganglia can be affected, or the putamen, caudate nucleus, thalamus may be involved in isolation.

- The white matter is the second most common site of damage, involving bilaterally symmetrical confluent lesions in the centrum semiovale and periventricular white matter.

- In comatose patients with bilateral symmetrical signal abnormalities of the globus pallidus, together with confluent white matter lesions, especially in the centrum semiovale and periventricular, CO poisoning should be suspected.

II.1.6. Mitochondrial Disease: Leigh's Syndrome (Figure 7,8):

- Subacute necrotising encephalomyelopathy or Leigh's syndrome is an inherited autosomal recessive defect in the enzyme pathway for respiratory metabolism.

- Infantile (less than 2 years) and juvenile forms have variable onset of psychomotor regression, weakness, seizures, dystonia and cerebellar dysfunction, leading to death from progressive respiratory failure.
MRI: symmetric areas of T2 prolongation in the basal ganglia, periaqueductal region, and cerebral peduncles, with putaminal involvement being a consistent feature.

When Leigh disease is suspected, MR spectroscopy (best performed with long echo times) may reveal the presence of abnormally high lactate levels in the basal ganglia, which together with elevated serum and CSF lactate levels supports the diagnosis.

II.2. Vascular causes:

II.2.1. Arterial Occlusion (Figure 9):

- Bilateral acute synchronous arterial infarctions of the thalamus are not uncommon, and are usually the result of occlusion of the rostral basilar artery.

- Thrombosis of the rostral basilar artery typically also causes acute infarction of the midbrain and portions of the temporal and occipital lobes fed by the posterior cerebral artery, or of portions of the cerebellum fed by other branches of the vertebrobasilar arterial system.

- A rare cause of bilaterally symmetric thalamic infarction is occlusion of the artery of Percheron, an anatomic variant of the posterior circulation.

- Clinical signs: agitation, obtundation or coma, memory dysfunctions, and various types of ocular and behavioral changes.

- Imaging: * Hyperintensity on T2-weighted MR images and restricted diffusion on diffusion-weighted images

* The causative steno-occlusive disease involving the basilar artery is often well depicted on MR angiograms

II.2.2. Deep Cerebral Venous Thrombosis (Figure 10):

- The deep venous system comprises the internal cerebral veins, vein of Galen and straight sinus and it drains the hemispheric white matter, diencephalon and deep nuclei.

- Risk factors: hypercoagulable state during pregnancy or puerperium, use of oral contraceptives, vasculitis, and intracranial or systemic infections

- Clinical signs: headache and non-specific clinical features, disturbances of consciousness, eyemovements, lethargy and long tract signs, and death.

- Imaging: * Indirect MRI findings:

  Bilateral thalamic lesions, often with involvement of the basal ganglia.
Swelling and T2 prolongation maybe accompanied by haemorrhagic complication, additional involvement of cerebellum, brainstem or cerebral cortex and hydrocephalus.

* Direct signs:

Direct signs of the thrombosed vein may be subtle on conventional neuroimaging, but MR venography or CT venography is diagnostic.

II.2.3. Ischemic Anoxia (Figure 11):

-The etiology of a severe hypoxic insult varies for older children and adults versus neonates.

- Anoxia generally occurs in the setting of trauma, near drowning, drug overdose, or cardiopulmonary arrest.

- Gray matter is more affected than white matter. The caudate and putamen are more affected than the thalami in children and adults, with the reverse true for neonates.

-MRI:

* Early MR findings of anoxia are subtle in the first 24 to 48 hours. During the acute phase (less than 24 hours), diffusion-weighted images show abnormal signal in the basal ganglia, cerebellum, and cortex.

* T1-weighted images generally show hypointensity in the basal ganglia and cortex in children and adults versus high signal in the globus pallidus, putamen, and thalamus in neonates.

* T2-weighted images are highly variable but generally reveal hyperintensity in the basal ganglia and cortex.

* Proton-density images may be more sensitive than T2-weighted images in the early period after injury.

* Single voxel proton MR spectroscopy can be used to detect high levels of lactate in the brain of asphyxiated neonates, which is indicative of a poor outcome.

II.3. Infectious causes:

II.3.1. Transmissible Prion Disease: Creutzfeldt-Jakob Disease (CJD) (Figure 12):

-Creutzfeldt-Jakob disease is a fatal dementing spongiform neurodegenerative disease.

- It is the most common human transmissible prion disease, and comprises 4 main subtypes: sporadic, familial, iatrogenic and variant CJD.
- Patients with sporadic CJD develop rapidly progressive dementia, myoclonus, and characteristic periodic sharp-wave complexes on electroencephalography.

- Some reports suggest that axonal transsynaptic spread of the prion agent from the caudate head to the anterior putamen and posteriorly may explain the early asymmetric and late bilateral symmetrical involvement.

-MRI: *bilateral symmetric abnormalities in the putamen, caudate head as well as involvement of the cerebral cortex.

* Although cortical lesions are subtle, they are often better detected on DW MR imaging.

* Restricted diffusion seen at diffusion weighted MR imaging is attributed to spongiform neuronal degeneration and is more sensitive than T2-weighted imaging findings in detecting CJD, especially for cortical lesions.

**II.4. Inflammatory causes:**

**II.4.1. Neuro-Behçet Disease (Figure 13):**

- Behçet disease is a multisystemic, recurrent inflammatory disorder of unknown cause.

- Clinical signs: eg, headache, dysarthria, cerebellar signs, sensory signs, personality change.

- The CNS is affected in 4%-49% of patients with Behçet disease, which has a predilection for men.

- The sites of lesions include the brainstem, basal ganglia and thalamus, and less commonly, the white matter of the cerebral hemispheres and cervicothoracic spinal cord.

-MRI: These lesions are:

* hyperintense on T2-weighted,

* hypointense on T1-weighted images,

* enhance after contrast material administration,

* are typically associated with vasogenic edema.

* They are isointense or slightly hyperintense on diffusion-weighted images.

**II.5. Tumoral causes:**
II.5.1. Primary Bilateral Thalamic Glioma (PBTG) (Figure 14):

- The thalamus is affected in 1%-1.5% of brain tumors, including secondary involvement by contiguous spread of adjacent lesions such as pineal germ cell tumors.

- Although PBTG is a low-grade astrocytoma (grade II), patients with PBTG, because of its deep location, have a very poor prognosis despite therapy.

- CT and MR imaging typically reveal a mass that symmetrically enlarges both sides of the thalamus.

- At MR imaging, PBTG appears hyperintense on T2-weighted images and isointense on T1-weighted images. Typically, these tumors do not enhance on postcontrast T1-weighted images.
Fig. 2: Wilson disease in a 35-year-old men with tremors and dystonia: (a)(b) and (c) T2-Flair-weighted image with bilaterally and symmetric areas of abnormal T2 prolongation in the thalamus (arrow), and lenticular nucleus. (d) T2-weighted axial MRI reveals the "face of the miniature panda" in the pontine tegmentum (arrowheads)

© - SFAX/TN

Fig. 3: Wernicke encephalopathy in a 32-year-old pregnant woman with impaired consciousness: Axial (a, b, c) and sagittal (d) T2-Flair-weighted image shows symmetric T2 prolongation in the medial thalamus, periaqueductal area, mamillary bodies, and tectal plate. diffusion-weighted (e) and ADC map(f) shows low ADC (late stage) in relation to irreversible damage.

© - SFAX/TN
**Fig. 4:** Acute Liver Disease: Axial T2-Flair-weighted (a), diffusion-weighted (b,c) and ADC map (d) shows bilaterally symmetric swelling, T2 prolongation, and restricted diffusion in the basal ganglia and cerebral cortex.

© - SFAX/TN

**Fig. 5:** Acute Liver Disease: MR spectroscopic detection of the combined toxic metabolite glutamate-glutamine at short echo times

© - SFAX/TN
**Fig. 6:** Carbon Monoxide Poisoning: Diffusion weighted magnetic resonance imaging (DWMRI) showed diffuse high signal intensity in both periventricular and deep white matter.

© - SFAX/TN
Fig. 8: Leigh’s Syndrome: MRI Brain Diffusion and Axial T2- weighted images: Bilateral symmetric T2 hyper intensity with faint high signal on diffusion involving putamen and caudate nuclei.

© Balaji Anvekar's Neuroradiology cases

Fig. 7: Leigh’s Syndrome: An upright Doublet of lactate at 1.3ppm on short TE of 35 ms with inversion at long TE of 144 ms on MRS.

© Balaji Anvekar's Neuroraduology cases
Fig. 9: Diffusion-weighted MRI of acute thalamic infarction in 2 patients. (A) Extensive tuberothalamic artery territory infarction on the left (right of image), as well as small lesions in the territory of the inferolateral arteries on the right. (B,C): Bilateral infarction in a teenager with patent foramen ovale: The infarcted region is in the tuberothalamic artery territory, although this pattern of bilateral stroke is more usually seen after paramedian artery infarcts.

© Massachusetts General Hospital

Fig. 10: Deep Cerebral Venous Thrombosis: T2WI(b) and FLAIR(a) MRI sequences showing abnormal hyperintensity in the thalamus bilaterally, extending into left basal ganglia and periventricular white matter Gradient echo image(c) showing susceptibility artefacts in the left thalamus and basal ganglia compatible with hemorrhagic component of venous infarct Sagittal T1w-Gado shows absent flow in the deep cerebral veins

© - SFAX/TN
Fig. 11: cardio-respiratory arrest in a 2-year-old baby: diffusion-weighted images (c,d) show abnormal signal and restricted diffusion in the basal ganglia, cerebellum, and cortex, whereas T2WI and FLAIR MRI sequences (a,b) are normal.

© - SFAX/TN
Fig. 12: Sample case of predominant striatal lesions in the early stage. Images were obtained at 3 (A and B) and 5 (C and D) months from the onset of symptoms. (A) FLAIR image shows changes, which are not as conspicuous as in B. (B) Striata appear hyperintense at diffusion-weighted imaging. Note that the anterior portion of the bilateral putamina (arrows) appears more hyperintense than does the posterior portion at diffusion-weighted imaging. C and D, Severe atrophy is depicted in both cerebral cortices and the caudate nuclei heads at FLAIR imaging (C) and diffusion-weighted imaging (D). Note that the putamina are entirely involved in C as compared with their appearance in B. Hyperintensity in the heads of the caudate nuclei appears less prominent; this appearance is associated with their volume loss and the dilatation of the frontal horns.

© AJNR 2002 23: 1164-1172
**Fig. 13:** Neuro-Behçet Disease: T2WI and FLAIR MRI sequences showing abnormal hyperintensity in the thalamus bilaterally, extending into left basal ganglia and cerebral peduncles, sparing the red nuclei without mass effect on adjacent structures

© - SFAX/TN

**Fig. 14:** Primary Bilateral Thalamic Glioma (PBTG): Coronal and axial T2-weighted MR images showed hyperintense bilateral thalamic and brainstem involvement with the posterior enlargement of lateral ventricles.

© Journal of Neurological Sciences (Turkish)
Fig. 1: Fahr Disease: Noncontrast CT scan shows bilaterally symmetric high-attenuation calcifications in the thalamus, caudate nuclei, putamina and globus pallidus,
Findings and procedure details

II.6. Other rares causes:

II.6.1. Metabolic and toxic causes:
* Nonketotic Hyperglycemia.
* Hypoglycemia.
* Osmotic Myelinolysis.
* Neurodegeneration with Brain Iron Accumulation.

II.6.2. Infectious causes:
* Flavivirus Encephalitis.
* Cerebral Toxoplasmosis.

II.6.3. Tumoral causes:
* Primary CNS Lymphoma.

II.6.4. Neurofibromatosis Type 1.
Conclusion

Systemic and metabolic abnormalities often involve the basal ganglia or thalamus on both sides, and careful assessment of brain abnormalities occurring simultaneously outside these structures is important. MR imaging, including T1-weighted imaging, diffusion-weighted imaging, MR angiography, MR venography, and MR spectroscopy, are often helpful in narrowing the differential diagnosis. Oftentimes, however, the diagnosis is not straightforward, and the correlation of typical imaging features with clinical and laboratory data can help make the correct diagnosis.
References


