MR Anatomy of Deep Brain Nuclei with Special Reference to Specific Diseases and Deep Brain Stimulation Localization

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Key words: MR imaging, deep brain stimulation, deep brain nuclei

SUMMARY – Diseases affecting the basal ganglia and deep brain structures vary widely in etiology and include metabolic, infectious, ischemic, and neurodegenerative conditions. Some neurologic diseases, such as Wernicke encephalopathy or pseudohypoparathyroidism, require specific treatments, which if unrecognized could lead to further complications. Other pathologies, such as hypertrophic olivary degeneration, if not properly diagnosed may be mistaken for a primary medullary neoplasm and create unnecessary concern. The deep brain structures are complex and can be difficult to distinguish on routine imaging. It is imperative that radiologists first understand the intrinsic anatomic relationships between the different basal ganglia nuclei and deep brain structures with magnetic resonance (MR) imaging. It is important to understand the “normal” MR signal characteristics, locations, and appearances of these structures. This is essential to recognizing diseases affecting the basal ganglia and deep brain structures, especially since most of these diseases result in symmetrical, and therefore less noticeable, abnormalities. It is also crucial that neurosurgeons correctly identify the deep brain nuclei presurgically for positioning deep brain stimulator leads, the most important being the subthalamic nucleus for Parkinson syndromes and the thalamic ventral intermediate nucleus for essential tremor. Radiologists will be able to better assist clinicians in diagnosis and treatment once they are able to accurately localize specific deep brain structures.

Introduction

This article reviews the normal MR anatomy of the deep brain with respect to intrinsic MR signal characteristics, shapes, appearances, and relationships to nearby nuclei and white matter tracts. The structures reviewed include the caudate, putamen, globus pallidus, thalamus, subthalamic nucleus, red nucleus, substantia nigra, pons, and medulla. The individual cranial nerve nuclei will not be discussed, as this will be beyond the scope of the review. Identification of the thalamic ventral intermediate nucleus and subthalamic nucleus will be discussed in further detail with regard to deep brain stimulation. We will then present many important neurologic diseases affecting deep brain structures, including Huntington's disease, conditions with basal ganglia calcification, hepatic encephalopathy, hypoxic ischemic encephalopathy, Creutz-

feldt-Jacob disease, Wernicke encephalopathy, hypertrophic olivary degeneration, Wallerian degeneration, Parkinson's disease, progressive supranuclear palsy, multisystem atrophy, and postoperative encephalopathy with choreoathetosis. Finally, we will briefly discuss the role and indications for deep brain stimulation.

Deep Brain Anatomy

Striatum

Two of the most easily identifiable nuclei of the deep brain are the caudate and putamen, which together form the striatum (Figure 1). The caudate nucleus is C-shaped and forms the floor of the frontal horn and body and the roof of the temporal horn of the lateral ventricle. The putamen is a curved structure bordered on

Figure 1 3 Tesla axial T2 MR image of a healthy subject shows the head of the caudate (C) and putamen (P), which together make up the striatum. They are both isointense to gray matter on T2-weighted imaging. With age, these nuclei demonstrate a gradual decrease in T2 signal, primarily due to iron deposition. The caudate and putamen are separated from one another by the anterior limb of the internal capsule, which can be seen as a relatively T2-hypointense linear structure, due to high myelin or lipid content. The globus pallidus (GP) lies medial to the putamen. The GP externa and interna, highlighted in red, are located laterally and medially, respectively. The globus pallidus is hypointense on T2 imaging, due to iron deposition.

its convex margin by the external capsule. The putamen is separated from the caudate by the anterior limb of the internal capsule.

The striatum is involved in the control of movement and communicates with the motor cortex, substantia nigra, subthalamic nucleus, and globus pallidus. Hence, pathologic processes of the striatum, or the nuclei with which they are in communication, can result in movement disorders. Atrophy of the striatum is seen in Huntington's disease and the Parkinsonian subtype of multiple system atrophy (MSA-P).

Globus Pallidus

The globus pallidus is located medial to the putamen and lateral to the posterior limb of the internal capsule (Figure 1). The globus pallidus and putamen together form a lens-shaped structure, referred to as the "lentiform nucleus". The globus pallidus demonstrates less signal intensity than the adjacent putamen on T2-weighted MR imaging due to iron deposition and is relatively T1-hyperintense due to myelination. The globus pallidus is divided into the globus pallidus interna (GPi) and globus pallidus externa (GPe), which are separated by the poorly visualized internal medullary lamina. The GPe is separated from the putamen by the also poorly visualized external medullary lamina. Similar to the striatum, the globus pallidus is responsible for

controlling fine motor movement and communicates with the striatum, substantia nigra, putamen, and thalamus. T1 hyperintensity of the globus pallidus can be seen in chronic hepatic encephalopathy due to deposition of paramagnetic manganese. Ischemic injury in the case of carbon monoxide poisoning results in T2 hyperintensity of the globus pallidus and possibly T1 hyperintensity if there is hemorrhage.

Thalamus

The thalamus is T2-isointense to gray matter and is located posterior to the interventricular foramen of Monro and medial to the posterior limb of the internal capsule (Figure 2A). The pair of thalami forms the borders of the third ventricle. The thalamus relays sensory and motor function between the cerebral cortex and spinal cord. It also regulates consciousness via the reticular activating system.

The thalamus is further divided into medial, lateral, and anterior groups of nuclei by the Y-shaped internal medullary lamina, which is not easily identifiable with routine MR imaging (Figure 2B). The ventral intermediate nucleus (VIM) is commonly targeted with deep brain stimulation (DBS) for the treatment of essential tremor (Figure 2C). The VIM is located laterally within the thalamus and is situated between the ventral lateral (VL) and the ventral posteromedial (VPM) nuclei¹.

Figure 2 A) 3 Tesla axial T2 MR image of a healthy subject shows the thalamus isointense to gray matter (highlighted in red) located posterior to the foramen of Monro. B) The individual thalamic nuclei are difficult to visualize, but the anterior nucleus and pulvinar can be identified by their exterior locations, anteriorly and posteriorly, respectively. C) Axial susceptibility-weighted imaging shows the ventral intermediate nucleus (VIM) located within the ventral thalamus just lateral to the red nucleus (RN) (outlined in red). Practical MR localization is by identifying the superiormost aspect of the hypointense red nucleus in axial section and tracking the small hypointense band (VIM nucleus) extending medial to lateral and slightly angulated posteriorly.

Figure 3 3 Tesla coronal T2 MR image of a healthy subject shows the hypointense subthalamic nucleus (STN) located superolateral to the hypointense substantia nigra (SN) and inferomedial to the internal capsule, which in turn is medial to the hypointense globus pallidus (GP). The substantia nigra demonstrates low T2 signal due to iron deposition².

Figure 4 A) 3 Tesla midline sagittal T1 MR image of a healthy subject shows the mammillary body (white arrow) located at the postero-inferior aspect of the hypothalamus. The mammillary bodies are part of the limbic pathway, which involves the hippocampus, fornix, mammillary body, mammillothalamic tract, anterior nucleus of the thalamus, and lastly the cingulate gyrus. B) Axial T2 image shows the pair of mammillary bodies anterior to the cerebral peduncles. C) Coronal T2 image shows the pair of mammillary bodies. Hippocampal and temporal cortical atrophy is assessed at the same coronal level as the mammillary bodies in Duara's visual rating system for Alzheimer's disease⁴.

Figure 5 A) 3 Tesla axial T2 MR image of a healthy subject shows the pair of hypointense red nuclei (red arrow) within the midbrain at the same level as the hypointense substantia nigra (white arrow). B) Axial susceptibility-weighted image shows the pair of red nuclei (red arrow) as hypointense structures, due to iron deposition. The substantia nigra pars reticulata (black arrow) is also hypointense due to iron deposition. The isointense pars compacta is located between the pars reticulata and red nucleus. One way to remember the relationship of structures, from medial to lateral, is "RCR (Royal College of Radiologists)." This stands for Red nucleus, pars Compacta, and pars Reticulata.

Figure 6 3 Tesla axial T2 MR images of a healthy subject show the pons at the level of the superior cerebellar peduncles (red arrow) (A) and at the level of the middle cerebellar peduncles (between red arrows) (B).

Subthalamic Nucleus

The subthalamic nucleus (STN) is a small, almond-shaped T2-hypointense structure within the rostral midbrain located lateral to the red nucleus, superior to the substantia nigra, and medial to the internal capsule (Figure 3). It is difficult to visually separate from

the T2-hypointense substantia nigra. However, the STN is reliably located between 9.7 and 9.9 mm lateral to midline at the level of the rostral cerebral peduncles³. The STN communicates with the GPi, GPe, substantia nigra, and the pedunculopontine nucleus (PPN). The substantia nigra and the GPe contribute to the latero-pallido-subthalamic system, which is also re-

Figure 7 3 Tesla axial T2 MR image at the level of the medulla. The red structure represents the shape and location of the inferior olivary nucleus.

ferred to as the “central pacemaker of the basal ganglia” and is thought to play an important role in Parkinson’s disease. The STN is a common target for deep brain stimulation (DBS) in the treatment of Parkinson syndromes.

Mammillary Bodies

The mammillary bodies are the pair of easily identifiable structures located at the posteroinferior border of the hypothalamus (Figure 4) and are necessary components of the limbic pathway, or Papez circuit. Mammillary body atrophy is a very important secondary finding of mesial temporal sclerosis, as it is connected to the hippocampal white matter through the fornix, via the limbic pathway. Lesions involving the mammillary body result in amnesic disorders, such as Wernicke-Korsakoff syndrome.

The mammillary body is also used as a landmark to standardize evaluation for mesial temporal atrophy in dementia. The hippocampus and perirhinal and entorhinal cortices are evaluated in the coronal plane perpendicular to the anterior commissure-posterior commissure plane. Greater atrophy correlates with Alzheimer’s disease, with a grade of 0 being normal and a grade of 4 being severely atrophied⁴.

Red Nuclei

The red nuclei are reliably recognized as a pair of round T2-hypointense structures located within the rostral midbrain (Figure 5). The reason for the low T2 and susceptibility-weighted imaging signal is because of its high iron content. The red nucleus is responsible for gross motor function via the rubrospinal tract.

It receives afferent projections from the contralateral cerebellar dentate nucleus and sends efferent projections to the ipsilateral inferior olivary nucleus. Lesions involving the red nucleus may result in hypertrophic olivary degeneration characterized by a movement disorder called “patalatal myclonus.”

Substantia Nigra

The substantia nigra is a small curvilinear structure located just anterolateral to the red nucleus and medial to the cerebral peduncle. It is composed of two nuclei, the pars compacta and the pars reticulata (Figure 5). The name substantia nigra, or “black substance”, is due to the predominance of dopaminergic neurons in the pars compacta, which produce dark melanin, thus giving a black appearance on gross specimen. The pars reticulata demonstrates low signal on T2- and susceptibility-weighted imaging due to its high iron content. The pars compacta, on the other hand, contains half the amount of iron and is, therefore, isointense to gray matter. The pars compacta supplies dopaminergic input to the striatum and globus pallidus, and its dysfunction is central to Parkinson’s disease and Parkinson plus syndromes. To support such a diagnosis, atrophy of the pars compacta greater than two standard deviations below the mean (or less than 2.4 mm in thickness) can be assessed with axial T2-weighted imaging⁵.

Pons

The pons is located between the midbrain and the medulla (Figure 6). The pons is made of a ventral (basilar) and a dorsal part. The

Figure 8 Huntington's disease. 1.5 Tesla axial T2 MR (A) and coronal T1 MR images (B) demonstrate severe atrophy of the caudate and putamen with resulting dilatation of the lateral ventricles in a "boxcar" appearance.

Figure 9 Pseudohypoparathyroidism. 1.5 Tesla axial post-contrast T1 MR image shows bilateral and symmetric hyperintensity within the caudate and putamen due to calcification.

Figure 10 Chronic hepatic encephalopathy. 1.5 Tesla axial T1 MR image shows bilateral and symmetric globus pallidus hyperintensity due to deposition of paramagnetic manganese.

Figure 11 A) 1.5 Tesla axial diffusion-weighted MR and (B) FLAIR MR images in an adult with severe hypoxic ischemic injury demonstrate bilateral hyperintensity within the caudate, putamen, globus pallidus, thalamus, as well as diffuse cortical involvement. C) 1.5 Tesla axial T1 MR image of an infant with hypoxic ischemic injury demonstrates increased signal within the bilateral putamen, globus pallidus, and thalamus (red arrows), greater than that of the posterior limb of the internal capsule (white arrow), indicating a less favorable prognosis¹¹.

dorsal portion of the pons forms the floor of the fourth ventricle and contains nuclei of the trigeminal nerve, the abducens nucleus, and the facial motor nucleus, as well as the medial lemniscus. The ventral part contains the vertically oriented corticospinal tracts as well as many horizontally oriented pontocerebellar fibers, which form the middle cerebellar peduncles. Multiple system atrophy of the cerebellar-predominant type (MSA-C) is characterized by atrophy of the pontocerebellar fibers and middle cerebellar peduncles.

Medulla

The medulla is located between the pons and the cervical spinal cord (Figure 7). The corticospinal tracts continue caudally through the medulla within the ventrally located pyramids. The inferior olivary nucleus, which lies posterolateral to the pyramid, is a thin lamina of gray matter containing neurons that send efferent projections to the contralateral cerebellar dentate nucleus via the inferior cerebellar peduncle and receives afferent projections from the ipsilateral red nucleus via the rubro-olivary tract. Lesions affecting the afferent projections to the inferior olivary nucleus can result in enlargement and T2 hyperintensity, which is termed hypertrophic olivary degeneration.

Specific Diseases Involving the Deep Brain Nuclei

Huntington's Disease

Huntington's disease is a neurodegenerative disorder, which is inherited in an autosomal dominant fashion. It is diagnosed genetically by an increased number of CAG triplet repeats within the Huntingtin gene, which is located on the short arm of chromosome 4 (4p16.3) and codes for the Huntingtin protein. A higher number of CAG triplet repeats results in mutation of the Huntingtin protein and correlates with earlier onset and greater severity of the disorder. With each generation, the number of genomic CAG triplet repeats increases, which is known as genetic anticipation. Huntington's disease results in an early and disproportionate loss of neurons within the striatum (Figure 8)⁶. The remaining caudate and putamen demonstrate increased apparent diffusion coefficient values (ADC) on MR imaging. The degree of striatal volume loss and increase in ADC correlate with the severity of the disease⁷.

Intracranial Calcification

Pseudohypoparathyroidism is a genetic disorder of end-organ dysfunctional response to normal parathyroid hormone with the most

Figure 12 Sporadic Creutzfeldt-Jakob disease (sCJD). 1.5 Tesla axial FLAIR MR image shows bilateral and symmetric hyperintensity within the pulvinar and dorsomedial nuclei of the thalamus giving a "hockey stick" appearance (outlined in red). The hockey stick is less intense than the hyperintense caudate and anterior putamen (red arrows), suggestive of sporadic CJD^{12,13}.

Figure 13 Wernicke encephalopathy. 1.5 Tesla axial FLAIR MR image shows hyperintensity within the hypothalamus, surrounding the third ventricle, the mammillary bodies, and the periaqueductal gray matter.

Figure 14 Hypertrophic olivary degeneration caused by a pontine cavernoma. A) 1.5 Tesla axial gradient recalled echo MR image shows a hemosiderin-laden cavernoma within the dorsal pons involving the left rubro-olivary pathway (red arrow). B) 1.5 Tesla axial FLAIR MR image of the same patient shows hyperintensity and hypertrophy of the left inferior olivary nucleus (red arrow).

Figure 15 3 Tesla coronal MR image of a healthy subject demonstrating the Guillain-Mollaret triangle connecting the dentate nucleus (DN) in the cerebellum, the contralateral red nucleus (RN) in the midbrain, and the contralateral inferior olivary nucleus (ION) in the medulla. The arrows denote afferent and efferent directions of the fibers connecting to the ION. Primary lesions are represented as blue or red bars; blue, red, and red/blue bars represent primary lesions resulting in ipsilateral, contralateral, and bilateral hypertrophic olivary degeneration (HOD), respectively. Primary lesions affecting the superior midbrain (1) result in ipsilateral HOD. Primary lesions affecting the inferior midbrain (2) result in bilateral HOD due to involvement of ipsilateral superior cerebellar peduncle (SCP) fibers (dashed green line) within the SCP. Primary lesions affecting the rubro-olivary tract in the pons (3) result in ipsilateral HOD. Primary lesions affecting the SCP (4) result in contralateral HOD¹⁵.

Figure 16 Wallerian degeneration secondary to remote right middle cerebral artery infarction. A) 1.5 Tesla axial T2 MR image shows right frontotemporal lobe encephalomalacia (red arrow) and ipsilateral corticospinal tract hyperintensity within the cerebral peduncle (yellow arrow). B) More caudal axial T2 image of the same patient shows hyperintensity of the corticospinal tract within the medullary pyramid (yellow arrow), which is medial to the inferior olivary nucleus (circled in red). The differential for T2 hyperintensity within the medulla includes hypertrophic olivary degeneration and Wallerian degeneration.

commonly described type being Albright's hereditary osteodystrophy. The characteristic MR imaging finding in pseudohypoparathyroidism is T1 hyperintensity within the caudate, putamen, and cerebellar dentate nucleus due to calcification (Figure 9)⁸. This is not pathognomonic, however, and can be seen in any con-

dition with intracranial calcification like Fahr syndrome, Fabry disease, and other calcium metabolism abnormalities. T1 hyperintensity of the thalamic pulvinar nucleus has been characteristically described for Fabry disease, but this can also occur with any calcium deposition.

Figure 17 Progressive supranuclear palsy. A) 1.5 Tesla midline sagittal T1 MR image shows the “Penguin” or “Hummingbird sign” due to flattening of the superior profile of the midbrain drawn from a point immediately posterior to the mammillary body, anteriorly, to the anterior aspect of the aqueduct, posteriorly. This flattening of the midbrain superior profile is caused by atrophy of the periaqueductal gray matter and pretectal area as well as neuronal loss within the Edinger-Westphal nucleus and nucleus interstitialis of Cajal¹⁹. B) 3 Tesla midline sagittal T1 MR image of a healthy subject demonstrating the normal convex superior profile of the midbrain.

Figure 18 Progressive supranuclear palsy. 1.5 Tesla axial susceptibility-weighted MR image shows severe posterolateral putaminal hypointensity (red arrows) similar to the deep veins (Grade 3) (white arrow). Practically, in susceptibility-weighted MR imaging, if signal intensity of these deep brain nuclei is similar to cerebrospinal fluid, this is grade 0. If there is severe hypointensity similar to the vein of Galen, then this is grade 3. Grades 1 and 2 denote mild and moderate hypointensity, respectively¹⁹.

Figure 19 Multiple system atrophy of the cerebellar-predominant subtype (MSA-C). 1.5 Tesla axial T2 MR image shows a classic “hot cross bun” appearance of the pons due to atrophy of the pontocerebellar fibers and middle cerebellar peduncles.

Figure 20 Deep brain stimulation (DBS) of a pediatric patient with postoperative encephalopathy with choreoathetosis. Postoperative encephalopathy with choreoathetosis, also known as “post-pump chorea,” is a movement disorder that occurs in children following cardiopulmonary bypass for heart surgery. The exact cause is unknown, but it is hypothesized to result from hypothermia during surgery. Medications, including antipsychotics and epileptics, are usually ineffective and DBS is occasionally helpful²³. 1.5 Tesla coronal T1 MR image (A) and axial T2 MR images (B) show placement of the deep brain stimulator leads within the bilateral globus pallidus interna (red arrows), just lateral to the internal capsule (highlighted in green).

Hepatic Encephalopathy

Hepatic encephalopathy presents as a neuropsychiatric spectrum of abnormalities, including dementia, gait instability, tremors, and Parkinsonian features. Acute hepatic encephalopathy occurs in acute liver failure and results in cerebral edema. This is due to increased ammonia uptake by astrocytes and conversion to glutamine resulting in an increased intracellular osmotic gradient. MRI findings include diffusely increased cortical T2 signal with sparing of the perirolandic and occipital regions⁹.

Chronic hepatic encephalopathy occurs in cirrhotics or patients receiving total parenteral nutrition. Ninety percent of chronic hepatic encephalopathy patients demonstrate T1-hyperintense globus pallidi due to paramagnetic manganese deposition (Figure 10). Manganese deposition is reversible, and hyperintense globus pallidi return to normal within approximately one year following liver transplant⁹.

Hypoxic Ischemic Injury

Hypoxic ischemic injury can occur at any age and manifests on imaging differently depending on the level of brain maturation. Mild to moderate hypoxic ischemic injury may result in border-zone watershed infarcts. Severe hypoxic ischemic injury results in symmetric cortical, thalamic, and basal ganglia diffusion restriction and T2/FLAIR hyperintensity (Figure 11). The most sensitive imaging sequence within the first 24 hours is diffusion-weighted imaging, which may normalize after one week. After 24 hours, T2/FLAIR hyperintensity becomes apparent¹⁰. In neonates, the posterior limb of the internal capsule is normally hyperintense relative to the basal ganglia due to myelination on T1-weighted imaging. In neonatal hypoxic ischemic injury, there is a less favorable prognosis if the T1 signal intensity of the posterior limb of the internal capsule is less than that of the posterolateral putamen (Figure 11C)¹¹.

Creutzfeldt-Jakob Disease

Creutzfeldt-Jakob Disease (CJD) is a rare type of rapidly progressive fatal dementia. Sporadic CJD (sCJD) is idiopathic, and variant CJD (vCJD) is thought to be caused by bovine spongiform encephalopathy. Diffusion-weighted and FLAIR sequences are most sensitive for detecting astrocytosis within the pulvinar ("pulvinar sign") and dorsomedial thalamic

nuclei (Figure 12). If FLAIR hyperintensity includes both the thalamic dorsomedial nuclei and pulvinar this is referred to as the "hockey stick sign". If the hockey stick is more intense than the caudate and anterior putamen this is diagnostic of vCJD. If the hockey stick is less intense than the caudate and anterior putamen, along with cortical hyperintensity, this is suggestive of sCJD^{12,13}.

Wernicke Encephalopathy

Wernicke encephalopathy is a reversible neurologic disorder caused by thiamine (vitamin B1) deficiency with alcohol abuse causing half of the cases. The classic triad of confusion, ocular dysfunction, and ataxia is present in only 38% of patients, therefore, making it difficult to diagnose clinically. Misdiagnosis can lead to irreversible Korsakoff psychosis. The most common MR finding is symmetric T2 hyperintensity of the medial thalami and regions surrounding the third ventricle (Figure 13). The next most common area of involvement is periaqueductal gray matter followed by the mammillary bodies. Contrast enhancement of the mammillary bodies is useful in differentiating alcoholic from nonalcoholic cases¹⁴.

Hypertrophic Olivary Degeneration

Lesions of the triangle of Guillain and Mollaret (dentato-rubro-olivary pathway) result in hypertrophy of the involved inferior olivary nucleus (ION) (Figure 14). It is important to recognize this entity to avoid misdiagnosis of a medullary infarct, demyelinating process, or neoplasm. The primary lesion is usually hemorrhage, cavernoma, diffuse axonal injury, infarct, demyelination, or tumor. The classic clinical presentation of hypertrophic olivary degeneration (HOD) is palatal myoclonus. Increased T2 signal of the ION follows the inciting lesion by one month and lasts up to four years. ION hypertrophy occurs by six months and lasts approximately four years, after which the ION may become atrophic¹⁵.

In order to understand the pathophysiology of HOD, it is important to become familiar with the anatomy of the triangle of Guillain and Mollaret (Figure 15). While olivary afferent pathway damage (rubro-olivary and dentato-rubral) results in HOD, olivary efferent pathway lesions (olivato-dentate), such as inferior cerebellar peduncle lesions, are less likely to cause HOD¹⁵.

Wallerian degeneration along the corticospinal tract could demonstrate T2 hyperintensity in the anteromedially located medullary pyramid and should be differentiated from HOD, which occurs more posterolaterally (Figure 16)¹⁶.

Parkinson's Disease

Parkinson's disease (PD) is a progressive neurodegenerative disease that is characterized by a resting tremor, bradykinesia, rigidity, and postural instability and is most commonly diagnosed in patients over 50 years of age. Loss of dopaminergic input to the striatum from the substantia nigra pars compacta is central to PD. The diagnosis is usually made clinically. However, neuroimaging is usually performed to exclude other treatable conditions prior to initiating pharmacologic therapy. In medically refractory cases, deep brain stimulation of the subthalamic nucleus or globus pallidus interna can be used¹⁷.

Progressive Supranuclear Palsy

Progressive supranuclear palsy (PSP) is a type of Parkinson plus syndrome. PSP is often clinically difficult to distinguish from Parkinson syndromes. The characteristic clinical features of PSP include supranuclear vertical gaze palsy, Parkinsonian features, postural and gait instability, and cognitive disturbance. On imaging, there is significant atrophy of the midbrain, superior colliculi, and superior cerebellar peduncles out of proportion to pontine atrophy (Figure 17)¹⁸. Three important imaging features to recognize can be remembered with the mnemonic "PSP": Pars compacta atrophy (seen with any Parkinson syndrome), Superior colliculus atrophy, and Periaqueductal gray matter hyperintensity.

Of all the Parkinson plus syndromes, PSP has the most iron deposition within the deep brain nuclei, including the putamen, red nucleus, substantia nigra pars reticulata, and cerebellar dentate nucleus (Figure 18)²⁰.

Multiple System Atrophy

Multiple system atrophy (MSA) is divided into three subtypes: MSA of the cerebellar-

predominant subtype (MSA-C), Parkinsonian subtype (MSA-P), and with autonomic failure (MSA-A), also known as "Shy Drager syndrome." MSA-C, also known as "olivopontocerebellar atrophy," is characterized by cerebellar ataxia. The characteristic imaging feature is atrophy of the pontocerebellar fibers and middle cerebellar peduncles resulting in the "hot cross bun" sign (Figure 19). MSA-P is characterized by extrapyramidal features and putaminal atrophy. On imaging, there is T2 hypointensity and atrophy of the putamen with a rim of T2 hyperintensity²¹.

Deep Brain Stimulation

Deep brain stimulation (DBS) is a type of functional neurosurgery using electrical leads, which was first introduced in the 1980s. The advantages of DBS are its reversibility, titratability, and adjustability in contrast to older, more invasive, neurosurgical approaches to psychiatric and movement disorders. DBS was first developed for the treatment of Parkinson's disease and essential tremor but is now used to treat multiple movement and psychiatric disorders (Figure 20)²².

Conclusion

The deep brain structures are a complex group of nuclei and white matter tracts that are sometimes difficult to identify on routine imaging.

Correct identification of specific nuclei can alter treatment, such as the case with typical and atypical Parkinson syndromes, other rare but important conditions like Creutzfeldt-Jakob disease, and conditions like hypertrophic olivary degeneration.

It is also crucial that neurosurgeons correctly identify these nuclei, presurgically, for positioning deep brain stimulator leads, the most important being the subthalamic nucleus for Parkinson syndromes and the thalamic ventral intermediate nucleus for essential tremor. Radiologists will be able to better assist clinicians in diagnosis and treatment once they are able to accurately localize specific deep brain structures.

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