Role of Magnetic Resonance Imaging in the Evaluation of Basal Ganglia Lesions

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ABSTRACT

Introduction: Radiologists will encounter bilaterally symmetrical abnormalities of the basal ganglia, usually in diffuse systemic and metabolic diseases. Study aimed to identify the spectrum of diseases involving basal ganglia disease and also to evaluate other sites of involvement in these patients.

Material and methods: 30 patients referred to our hospital with suspicion of basal ganglia lesions subjected to MR examinations by 3TMR system using a 32-channel phased array head coil.

Results: Most common etio-pathologies affecting basal ganglia are metabolic diseases (67%), Japanese encephalitis and dengue encephalitis (17%), Neoplasm’s Glioma and DNET (10%) and degenerative NBIA and Huntington’s disease (7%) respectively. Among metabolic diseases Hypoxic ischemic encephalopathy (10%) and posterior reversible encephalopathy syndrome (10%) diagnosed respectively. Among the Thirty patients with basal ganglia lesions, bilateral symmetrical involvement in 22 cases (73%), unilateral involvement in 5 cases (17%) and bilateral asymmetrical involvement in 3 cases (10%). There are 5 cases with Caudate +Putamen (Dorsal striatum), 4 cases with Putamen+globus pallidus (lentiform nucleus) and 7 cases with thentire basal ganglia (Corpus striatum) involvement in metabolic disease patients.

Conclusion: MR imaging has dramatically improved the ability to visualize deep grey structures of basal ganglia and spectrum.

Key words: Basal Ganglia Lesions, Magnetic Resonance imaging, Huntington’s Disease.

INTRODUCTION

The deep grey matter structures of the basal ganglia comprise the caudate nucleus, putamen and globus pallidus. Computed tomography (CT) and magnetic resonance (MR) imaging have dramatically improved the ability to visualize the deep gray structures of the basal ganglia. 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. MRI allows detailed visualization of the morphology, signal intensity and metabolic content of the DGM nuclei, together with visualization of normal cortical development and normal myelination of the white matter. The basal ganglia 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 that alters cerebral metabolism, which can lead to selective damage to the basal ganglia. However, damage to the deep grey matter nuclei may be visualised either as basal ganglia lesions in isolation or as part of more generalised brain damage also involving other grey or white matter structures. Among the many causes of basal ganglia damage, bilateral symmetrical lesions typically are caused by diffuse systemic or metabolic conditions. Often, the cause (for example hypoxic ischemic encephalopathy in the context of cardiac arrest) is readily evident, but sometimes, the radiologist is faced with situations of bilateral symmetrical lesions in the basal ganglia for initial diagnosis (for example, coma of unknown cause) before the results of relevant investigations are known. There is considerable variation and overlap in both the clinical and radiologic features of abnormalities affecting the deep gray matter nuclei. Hence, no classification scheme is foolproof. The systematic approach to this uncommon group of systemic and metabolic conditions is often challenging, but assessment of all the neuroimaging findings (not merely of the basal ganglia but other features) is an essential component for final diagnosis and sometimes prognosis. The neuroimaging diagnosis is influenced not only by detection of specific MR imaging features such as restricted diffusion and the presence of haemorrhage, but also by detection of abnormalities involving other parts of the brain,
especially the cerebral cortex, brainstem, and white matter. Judicious use of confirmatory neuroimaging investigations, especially diffusion-weighted imaging, MR angiography, MR venography, and MR spectroscopy during the same examination, may help improve characterization of these abnormalities and help narrow the differential diagnosis. Recognition and correct evaluation of basal ganglia abnormalities, together with a proper clinical history and laboratory findings, may enable the identification of spectrum of disease entities and lead to earlier diagnosis. Reaching a diagnosis in the early stages of acute diseases in many patients is crucial for instigating prompt specific therapy leading to a favourable outcome. 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 aim of the study was to evaluate the spectrum of pathologic conditions of basal ganglia involvement (Excluding vascular pathologies), and the radiological assessment of these conditions with clinical correlation.

MATERIAL AND METHODS

Patients attending the department of Radio-diagnosis, Narayana Medical College, Nellore were the main source of data for this study. The study was approved by the institutional ethical committee and study details were explained to all the cases or relatives and consent was obtained. All patients referred to the department of Radiology with clinical suspicion of basal ganglia (deep grey matter) lesions in a period of 2yrs from SEP 2015 to SEP 2017 will be subjected for study. Total 30 patients with basal ganglia involvement were included based on inclusion criteria.

Inclusion Criteria: Patients with extrapyramidal symptoms, patients with suspected metabolic encephalopathy, all patients with incidentally diagnosed basal ganglia lesions by CT, follow up patients of basal ganglia lesions, cases of all age groups irrespective of sex.

Exclusion Criteria: Vascular pathologies of basal ganglia (Infarcts and haemorrhages), Patient having history of claustrophobia, Patient having history of metallic implants insertion, cardiac pacemakers and metallic foreign body in situ.

Equipment: The MR examinations were performed on all patients who met inclusion criteria at a 3T whole-body MR system (DISCOVERY 750W 3T MRI GE Healthcare, Chicago, Illinois) using a 32-channel phased array head coil.

Sequences: Conventional spin echo sequences, axial T1, T2 and FLAIR; Coronal FLAIR, DWI; SWAN and Post-contrast T1+C if required. Special sequences such as Single voxel spectroscopy was performed at TE of 33 and 144 msec were used as and when required.

STATISTICAL ANALYSIS

Statistical analysis was performed using percentages and proportions with the help of microsoft office 2015.

RESULTS

Table 1: Distribution of primary basal ganglia nuclei involvement among metabolic disease.

<table>
<thead>
<tr>
<th>Pathology</th>
<th>Caudate only</th>
<th>Caudate + Putamen</th>
<th>Putamen only</th>
<th>Putamen + GP</th>
<th>GP only</th>
<th>Corpus striatum</th>
</tr>
</thead>
<tbody>
<tr>
<td>HIE</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>_</td>
<td>2</td>
</tr>
<tr>
<td>Leigh’s syndrome</td>
<td>_</td>
<td>1</td>
<td>_</td>
<td>1</td>
<td>_</td>
<td>4</td>
</tr>
<tr>
<td>Hypoglycemic encephalopathy</td>
<td>_</td>
<td>_</td>
<td>_</td>
<td>_</td>
<td>_</td>
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<tr>
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<td>1</td>
<td>_</td>
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<tr>
<td>MSUD</td>
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<td>_</td>
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<td>_</td>
</tr>
<tr>
<td>Osmotic myelinolysis</td>
<td>_</td>
<td>1</td>
<td>_</td>
<td>_</td>
<td>_</td>
<td>_</td>
</tr>
<tr>
<td>Wilson’s disease</td>
<td>_</td>
<td>1</td>
<td>_</td>
<td>_</td>
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<td>_</td>
</tr>
</tbody>
</table>

Table 2: Distribution of primary basal ganglia involvement along with other areas of neuroparenchymal involvement in various metabolic diseases.

<table>
<thead>
<tr>
<th>Pathology</th>
<th>Basal ganglia + Thalamus</th>
<th>Basal ganglia + Cortex</th>
<th>Basal ganglia + White matter</th>
<th>Basal ganglia + Brainstem</th>
<th>Basal ganglia + Cortex + White matter</th>
</tr>
</thead>
<tbody>
<tr>
<td>HIE</td>
<td>5</td>
<td>6</td>
<td>2</td>
<td>-</td>
<td>2</td>
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<tr>
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<td>_</td>
<td>_</td>
<td>5</td>
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</tr>
<tr>
<td>Hypoglycemic encephalopathy</td>
<td>_</td>
<td>2</td>
<td>2</td>
<td>_</td>
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<tr>
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<td>Wilson’s disease</td>
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</table>
involvement. Thirty patients were evaluated, whose age
group ranged from 1 to >60 years. The highest incidence of
basal ganglia lesions were found in 21-30 years age group
accounting for 23% of cases and least was seen in age group
of 41-50 years constituting 3%. Thirty patients were evaluated
of which 18 (60%) were males and 12 (40%) were females.

Out of the 30 patients who were evaluated, Metabolic
diseases accounts for 20 cases (67%) and is the most common
pathology followed by infectious diseases representing 5
cases (17%), Neoplasm’s accounting for 3 cases (10%) and
degenerative diseases involving 2 cases (7%).

Among metabolic diseases, there are 6 cases each with
Hypoxic ischemic encephalopathy and Leigh’s syndrome,
2 cases each with hypoglycemic encephalopathy, posterior
reversible encephalopathy syndrome and 1 case each with
Maple syrup urine disease, Hepatic encephalopathy, Wilson’s
disease, Extrapyramidal myelinolysis.

Among thirty patients with basal ganglia lesions, seizures are
the most common presentation followed by altered mental
state especially in acute cases like metabolic and infectious
diseases. Extrapyramidal symptoms (Gait disturbances,
abnormal dancing movement, hypertonia, dystonia, rigidity
etc.) are the common presentation in chronic cases like
degenerative and neoplasm’s.

Incidence of 20 cases diagnosed with metabolic, 5 cases of
infectious, 3 cases of neoplasms, and cases of degenerative diseases respectively. 22 cases identified to be bilateral symmetrical side of pathology in basal ganglia lesions. Among Thirty patients, 24 (80%) cases showed diffusion restriction (partial/complete) and 6 (20%) cases showed no restriction. Out of Thirty patients evaluated, metabolic diseases are seen in 20 cases (67%). Among the 20 cases Males were 11 and Females were 9. Bilateral symmetrical involvement of basal ganglia is seen in 19 cases (95%) and unilateral involvement is noted in 1 case (5%).

Most common primary basal ganglia nuclei involved was caudate + lentiform nucleus (Corpus striatum) in 7 cases (35%), followed by Caudate + Putamen in 5 cases (25%), Putamen + GP (Lentiform nucleus) in 4 cases (20%), only GP in 2 cases (10%), only putamen in 1 case (5%) and lastly only caudate in 1 case (5%) (Table 1) (Figure 1). Involvement of most common site of neuroparenchyma in addition to basal ganglia includes BG + Thalamus in 9 cases (45%), followed by BG + Cortex in 10 cases (50%), BG + Cortex + White matter in 5 cases (25%), BG + White matter in 6 cases (30%) and finally BG + Brainstem in 8 cases (40%) (Table 2) (Figure 2).

**DISCUSSION**

Magnetic resonance imaging is a non-invasive, multiplanar and highly accurate method with better inherent contrast that demonstrates the lesion accurately. MRI provides an accurate assessment of the brain changes in various spectrum of basal ganglia lesion. This was a Hospital based prospective study done in the Department of Radio diagnosis and Imaging, Narayana medical college and hospital, aimed at studying the MR appearances/patterns in various spectrums of Basal ganglia lesions. In our study of MR imaging of basal ganglia lesions, we evaluated 30 patients. Out of the 30 patients who were evaluated, Metabolic diseases accounts for 20 cases (67%) and is the most common pathology followed by infectious diseases representing 5 cases (17%), Neoplasms accounting for 3 cases (10%) and Degenerative diseases involving 2 cases (7%). Finelli et al, in his case series study stated, metabolic conditions were the most common cause of acute bilateral symmetric DGMN lesions. Among metabolic diseases HIE (30%) and Leigh’s syndrome (30%) are the most common entities followed by hypoglycemic encephalopathy (10%) and PRES (10%). Hepatic encephalopathy, Wilson disease, Maple syrup urine disease and extraontine myelinolysis are other fewer pathologies accounting for rest of the metabolic diseases (20%). Viral encephalitis like Japanese encephalitis and dengue encephalitis are the most common pathologies under infectious diseases. Glioma and DNET are the pathologies included under Neoplasms’.

Among metabolic diseases HIE (30%) and Leigh’s syndrome (30%) are the most common entities followed by hypoglycemic encephalopathy (10%) and PRES (10%). Hepatic encephalopathy, Wilson disease, Maple syrup urine disease and extraontine myelinolysis are other fewer pathologies accounting for rest of the metabolic diseases (20%). Viral encephalitis like Japanese encephalitis and dengue encephalitis are the most common pathologies under infectious diseases. Glioma and DNET are the pathologies included under Neoplasms’. NBIA and Huntington’s disease are the disease entities which represents the degenerative diseases. Among thirty patients with basal ganglia lesions, seizures are the most common presentation followed by altered mental state especially in acute cases like metabolic and infectious diseases. Extrapyramidal symptoms (Gait disturbances, abnormal dancing movement, hypertonia, dystonia, rigidity etc.) are the common presentation in chronic cases like degenerative and neoplasm’s. Among the Thirty patients with basal ganglia lesions, bilateral symmetrical involvement noted in 22 cases (73%), unilateral involvement noted in 5 cases (17%) and bilateral asymmetrical involvement noted in 3 cases (10%).

AmongThirty patients who were evaluated; 24 (80%) cases showed diffusion restriction (partial/complete) and 6 (20%) cases showed no restriction. Out of thirty patients, metabolic diseases are seen in 20 cases (67%). Among the 20 cases Males were 11 and Females were 9.

Bilateral symmetrical involvement of basal ganglia is seen in 19 cases (95%) and unilateral involvement is noted in 1 case (5%). Most common primary basal ganglia nuclei involved was caudate + lentiform nucleus (Corpus striatum) in 7 cases (35%), followed by Caudate + Putamen in 5 cases (25%), Putamen + GP (Lentiform nucleus) in 4 cases (20%), only GP in 2 cases (10%), only putamen in 1 case (5%) and lastly only caudate in 1 case (5%) (Table 1) (Figure 1). Involvement of most common site of neuroparenchyma in addition to basal ganglia includes BG + Thalamus in 9 cases (45%), followed by BG + Cortex in 10 cases (50%), BG + Cortex + White matter in 5 cases (25%), BG + White matter in 6 cases (30%) and finally BG + Brainstem in 8 cases (40%) (Table 2) (Figure 2).

Involvement of most common site of neuroparenchyma in addition to basal ganglia includes BG + Thalamus in 9 cases (45%), followed by BG + Cortex in 10 cases (50%), BG + Cortex + White matter in 5 cases (25%), BG + White matter in 6 cases (30%) and finally BG + Brainstem in 8 cases (40%). Diffusion restriction was noted in 17 cases (85%) and in 3 cases (15%) no diffusion restriction is seen. Among 20 cases, various pathologies constitute the metabolic diseases. The most common entity among the metabolic diseases are HIE (6 cases, 30%) and Leigh’s syndrome (6 cases, 30%). HIE is the most common metabolic disease seen especially in adults.

Cardiac arrest was the mechanism of injury in the majority of the patients of our study population (5 cases). Cardiac causes accounted for 16 out of 39 cases of hypoxic-ischemic insult in the study by Choi et al, and 14 out of 22 cases in the study by Topcuoglu et al.

Wu et al, and Arbelaez et al, were focused only on patients with cardiac arrest as the cause of hypoxic-ischemic insult. Involvement of cerebral cortex (6 cases, 100%) was noted in association with basal ganglia noted in all cases. Thalamus involvement in addition to basal ganglia involvement is seen in 4 cases (67%). Hippocampal involvement in association with basal ganglia involvement noted in 3 cases (50%).

Diffuse cortical and deep grey matter pattern with periorbital involvement was the most frequent pattern across the major mechanism in a study by Eluvathingal Muttiikkalet al. Choi et al, in their study of 39 patients categorized the brain injuries into four patterns: normal, isolated cortical injury, isolated deep grey nuclei injury, and mixed injuries (cortex and deep grey).

Wu et al, concluded that whole-brain median ADC was a predictor of poor outcome. Leigh’s syndrome is next common entity among metabolic diseases which is exclusively seen in pediatric population especially neonates presenting with complaints of not accepting feeds, decreased activity, neuronal regression and seizures.

Medulla was normal without any hypertrophic olivary degeneration in our study. Brainstem and basal ganglia...
in six cases; the cerebellum, mesencephalon,pons, and supratentorial area in five cases; and the thalamus in four cases, respectively.

Felber et al.,21 demonstrated elevation of lactic acid, and presence of a peak at 0.9–1.0 ppm, belonging to methyl resonance of branched chain amino acids.

Wilson’s disease is another rare entity accounting for 1 case (5%). Paediatric female child who has hepatic failure presented with psychomotor illness and movement disorder. Laboratory investigation revealed copper traces in urine. On MRI evaluation, there is bilateral symmetrical T2 and FLAIR hyper intensity involving lentiform nucleus, thalamus, midbrain and pontine tegmentum without diffusion restriction.

T.J. Kim, I.O. Kim et al.,22 evaluated a cohort of pediatric Wilson disease patients in whom, the lentiform nuclei are involved most often, followed by the thalami, pons, midbrain, superior and middle cerebellar peduncles, and cerebellar nuclei.

Bilateral asymmetrical involvement was seen in 3 cases (60%), unilateral involvement and bilateral symmetrical involvement noted each in 1 case (20%). Diffusion restriction is seen in all cases (100%). Thalamus involvement is seen in all cases (100%) with associated haemorrhage was seen in 2 cases (40%). Basal cisterns, ventricles and meningeal enhancement are normal in all cases. Serology of 2 patients among encephalitis picture is positive for Dengue virus. The findings in rest of the 4 patients of our study reveals clinical features of encephalitis with negative dengue serology test and MRI imaging pattern shows deep grey matter and thalamic involvement which suggest the probability of JE and is considered in these 4 patients. J. Kalita et al.,21 evaluated forty two patients with JE. Thalamic lesions on CT and/or MRI combined had sensitivity 23% (95% confidence interval 12.9–33.1%), specificity 100%, positive predictive value 100% and negative predictive value 42.1% (95% confidence interval 30.2–53.8%) for a diagnosis of JE in a cohort of 75 patients with suspected encephalitis in a study by N. M. Dung and Lance Turtle etal.,(2009)23.

U.K. Misra, J. Kalita, et.al. evaluated 88 consecutive viral encephalitis patients, of them 22 patients had JE, 9 had dengue, 8 had herpes simplex encephalitis (HSE), 2 had Epstein-Barr virus encephalitis (EBVE) and 47 had non-specific encephalitis.

Sanjeev Kumar etal.,26 evaluated twenty-one serologically confirmed patients of dengue with altered sensorium who underwent MRI in them was 20. MRI was abnormal in 9 (45%).

SoniSR Das et al.,27 evaluated magnetic resonance imaging (MRI) findings of 3 patients with dengue fever diagnosed by positive dengue NS1 antigen revealed nonspecific imaging features of dengue encephalitis in two cases and dengue meningoencephalitis in one case in the form of diffuse cerebral oedema, bilateral symmetrical FLAIR and T2 hyper intensities in thalami, pons, and medulla with heterogenous or peripheral enhancement.

José M. García-Santos etal., described among brain tumours, those arising from the deep brain are rare. In many cases they
are low-grade astrocytomas followed by germ cell tumours. Wei Fu et al. retrospectively analyzed 35 children with PBGRT from December 2011 to December 2015. Most common were 15 astrocytomas and 11 germ cell tumours (GCT). DNET (Dyssembryoplastic neuroepithelial tumour) is the other neoplasm seen in our study. Young adult patient presented with recurrent history of partial seizures. No obvious perilesional edema noted. Associated cortical dysplasia or cortical tubers are not seen in our study. On T1+ C contrast study, no obvious enhancement of lesion seen. Clinico-radiologic criteria for the diagnosis of DNET are seen in our case.

These findings were very much similar to our study result of DNET. Degenerative diseases are rare ones involving basal ganglia accounting for 2 cases (7%) in my study. These diseases are confined only to basal ganglia nuclei without involvement of rest of the neuroparenchyma. Bilateral symmetrical involvement without obvious diffusion restriction is seen. Among the two cases, first one was NBIA (Previously known as Hallevordan Spatz disease) was seen in a 25 yrs old male presenting with movement disorder. On imaging, there was bilateral symmetrical globus pallidus involvement showing T1, T2 and SWAN hyper intensity with surrounding hypo intensity appearing as “Eye of Tiger” sign without diffusion restriction.

These findings were very much similar to our study result in NBIA. Second degenerative disease included in my study was Huntington’s disease, seen in a 40 yrs old male presenting with involuntary dancing movements (chorea). On evaluation, exclusive bilateral caudate and putamen (Caudate + Putamen) nucleus involvement noted in the form of atrophy. On axial images frontal horn of bilateral lateral ventricles appears as “Box Car” configuration.

CONCLUSION
Magnetic Resonance Imaging has dramatically improved the ability to visualise deep grey structures of basal ganglia and spectrum of diseases involving it. Basal ganglia has a characteristic appearance on MR sequences and abnormalities in their appearance have a characteristic pattern which helps in narrowing the differential diagnosis. Magnetic resonance imaging can contribute greatly to arriving at correct diagnosis with available clinical and laboratory data. Magnetic resonance imaging is useful in monitoring disease progression. It also has a prognostic implication.

REFERENCES