

Subcortical Syndromes: Neuropsychological Perspective

Mukesh Kumar Garg* & Preeti Sharma**

*Consultant Neurosurgeon, Ananta Institute of Medical Sciences, Udaipur

**Counselor, Academy of Well-being, Udaipur & Research Scholar, Pacific University, Udaipur

Abstract

Subcortical syndromes are the neurological disorders which are associated with the dysfunctioning or pathological functions of subcortical regions of the brain. The important subcortical regions of the brain are basal ganglia, thalamus, hypothalamus, and limbic system. Some of the most common subcortical syndromes include Parkinson's disease, Huntington's disease etc. The symptoms of these disorders include impairments in cognition, affect, and behavior. Genetic and biochemical factors are implicated in these disorders. The current paper will highlight the nature, types, symptoms and neuropsychological deficits in subcortical syndromes.

Key words: Subcortical regions, subcortical syndromes, neuropsychology

INTRODUCTION

Sub cortical nuclei are situated in the region just below the cerebral cortex. Subcortical structures primarily involve.

- Basal ganglia
- Limbic system
- Thalamus and
- Cerebellum

Basal Ganglia

Basal ganglia are large subcritical nuclear masses situated within each cerebral hemisphere. Basal ganglia consist of series of the subcortical structures, viz.

- Caudate nucleus
- Putamen
- Globus pallidus

These structures are closely interconnected with the substantia nigra in the midbrain. Basal nuclei are connected with many different regions of the nervous system by a very complex number of neurons.

Primary function of the basal ganglia is the motor control including

- Somatic motor function – control external body parts
- Oculomotor system – control of eye of movements and also involved in
- Executive functions – high level planning and control of motor skills.

The basal nuclei receive afferent information from most of the cerebral cortex, the thalamus sub thalamus and the information is integrated within the basal ganglia and influences the activities of the motor areas of the cerebra cortex or other motor

centers in the brain stem. Thus, the basal nuclei can control motor movements by influencing cerebral cortex, rather than through direct pathways to the spinal cord and brain stem. These areas, though clearly involved in motor control are not directly involved in control of muscular activities through spinal cord. There are no direct connections from the basal ganglia to the spinal cord. Rather, these areas have direct connections (or connections through thalamus) to motor cortex. So, they are part of the cortico- subcortical motor loop, but they do not provide the final connections to the spinal cord, destruction in any of these connections will result in motor/movement disorders.

Limbic System

Brain structures that are considered as part of the limbic system:

- Cingulate gyrus – a large region superior to the corpus callosum
- Hypothalamus
- Anterior thalamic nuclei (have shown to be involved in emotional responses)
- Amygdala
- Hippocampus
- Orbitofrontal cortex
- Parts of the basal ganglia

The areas which are called parts of the limbic system, are connected to each other, but they are also connected to other brain areas. The primary functional purpose is emotional responses (including emotional memory). The limbic system is also associated with the olfactory bulb, suggesting a structural explanation for the close relationship between smell and emotional responses.

Hypothalamus

The hypothalamus is highly involved in all aspects of endocrine, hormonal, visceral and autonomic functions. It is the control center for the control center for the activities such as eating, drinking, and the experience of pleasure, rage, and aversion. Hypothalamus is sexually dimorphic. Lateral and ventromedial hypothalamic nuclei play an important role in the control of the autonomic nervous system. These nuclei exert counterbalancing effect on each other for e.g. medial hypothalamus controls parasympathetic activities (reduction on heart rate, blood pressure) where as the lateral hypothalamus mediates sympathetic activity (increasing heart rate, blood pressure). Thus, these

nuclei regulate homeostasis with the help of the neurotransmitter acetylcholine. Maintains normal bodily functions such as the heart rate, digestion, thirst and sexual functioning. Hypothalamus controls the production, and release of many hormones, which regulate ANS function.

Amygdala

The amygdaloid nuclear complex is a gray mass in the dorsomedial part of the temporal lobe which underlies the uncus. This complex lies dorsal to the hippocampus and rostral to the tip of the inferior horn of the lateral ventricle. Lesions to the amygdala produce disturbances of emotional behavior. Le Daux has done significant work on the role of amygdala in emotional behavior. Hyper sexuality has been found as a prominent feature in some experimental studies. Bilateral ablations of amygdala result in hyperphagia, or in hypophagia.

Hippocampus

Hippocampus is a different structure than most of the cortex. It is not a part of neo cortex and has only 3-4 cell layers. Hippocampus is closely associated with the dentate gyrus. Parahippocampal gyrus and entorhinal cortex provide inputs to dentate gyrus and hippocampus. Most hippocampal connections are through the fornix. Some of these connections cross into the opposite hemisphere. Most connections however, stay within the same hemisphere and go to thalamus, through the mammillary bodies and hypothalamus. Hypothalamus is associated with learning and memory encoding and also involved in emotional processing particularly the anterior regions. The link between hippocampus and memory has been solidified with the study of H.M. who presented with the seizure disorder. With the removal of both hippocampuses (one in each hemisphere), explicit memory deficits were seen. However, implicit learning was restored.

Thalamus

Thalamus is part of the diencephalon (along with hypothalamus) and it is centrally located in the brain. It is often known as the relay station of the brain. It transfers all sensory receptors information except olfaction to higher brain centers. Thalamus is interconnected through a series of reciprocal (i.e., two way) connections to many perceptual areas. Almost all sensory inputs to the brain pass first through the thalamus. Pulvinar nucleus and reticular nucleus of thalamus play an important role in attention.

Cerebellum

The cerebellum is part of the brain. It lies under the cerebrum, towards the back, behind the brainstem and above the brainstem. Structurally, the cerebellum consists of

- A superficial gray mantle, the cerebellar cortex.
- An internal white mass, the medullar substance and
- Four pairs of intrinsic nuclei embedded in the white matter (dentate, emboli form globose and fastigius nuclei)

The traditional view on the functional role of cerebellum has been predominantly associated with motor control the adjustment of muscular tone the coordination of skilled voluntary movements and the regulation of posture and gait. Role of cerebellum in cognitive functions can be understood with reference to its connection with other areas of cortex. Cerebellum has been found to be involved in wide variety of cognitive functions such as intelligence, attention, motor functions, language, visuospatial abilities, learning and memory. Cerebellum also mediates executive functions such as set shifting, planning, fluency and working memory.

Damage to any of subcortical structures or malfunctioning in the subcortical areas is closely associated with diseases known as subcortical syndromes. Some of the diseases of the subcortical regions are:

Parkinson's Disease (PD)

PD is one of the most common chronic neurological disorders. Movement disturbances are the hallmark of Parkinson disease. Primary motor symptoms include akinesia, bradykinesia, rigidity and loss of associated movements, tremor and neuro-ophthalmic abnormalities.

Neuropathological studies suggest neuronal loss in the substantia nigra and other brainstem areas. And nonverbal memory for both immediate and delayed recall, visuospatial and visuo constructive skills. Executive functions are most frequently cited deficits. Depression anxiety and apathy are common psychiatric symptoms associated with PD.

Other Parkinsonian Syndromes

Striatonigral degeneration and PD are clinically similar, however tremors and dementia is not a prominent feature. Main clinical feature is in responsiveness to levodopa a drug commonly used to treat PD. Studies involving postmortem examination have shown neuronal loss in the putamen and caudate nuclei. Neuropsychological deficits include impairment in visuo spatial functioning, speech production, sentence repetition and abstract reasoning.

Idiopathic Calcification of Basal Ganglia (ICBG)

Excessive calcium deposits are found in globus pallidus, putamen, thalamus, and cerebella white matter. There is an early onset pattern occurs in

the second to fourth decade of life and initially as schizophrenia type psychosis. The late onset type ranges between ages 40 and 60 and marked by dementia and movement disorder, ICBG has characteristic subcortical features. Deficits include poor attention and concentration impaired abstraction and poor recent memory.

Wilson's Disease

Wilson's disease is hereditary in nature. Symptoms usually appear between the ages of 6 and 20 years but can begin as late as age 40. The most characteristic sign is the Kayser-Fleischer ring—a rusty brown ring around the cornea of the eye that can be seen only through an eye examination. The liver of a person who has Wilson's disease does not release copper into bile as it should. As the intestines absorb copper from food, the copper builds up in the liver and injures liver tissue. Eventually, the damage causes the liver to release the copper directly into the blood stream which carries the copper throughout the body. The copper build up leads to damage in the kidneys, brain and eyes. If not treated, Wilson's disease can cause severe brain damage, liver failure, and death

Huntington's Disease (HD)

HD is a progressive movement disorder. The main features of the disease are chorea the occurrence of rapid irregular and arrhythmic complex involuntary movements athetosis, dystonic motor restlessness, tremor, and myoclonus. It is a genetic condition, inherited as an autosomal dominant trait with complete lifetime penetrance.

The neuropathological feature is bilateral atrophy of the striatum. Neuronal depletion in the nuclei of the caudate nucleus and the putamen results in reduced neostriatal volume.

The pattern of cognitive impairment is described within the concept of "subcortical dementia" characterized by mental slowness, forgetfulness, impaired ability to manipulate acquired knowledge and affective changes. Behavioral changes are characterized by irritability impulsiveness depression or aggressiveness. The neuropsychological deficits are seen with regard to visual spatial cognition (pattern and spatial working memory spatial span and planning) impaired verbal fluency sequences generation, immediate recall and executive functions.

Gilles de la Tourette's Syndrome (GTS)

GTS presents with multiple motor tics and one or more vocal tics. Etiology involves a genetic basis as of single autosomal gene with varying penetrance. It has been hypothesized that an imbalance of the neurotransmitter dopamine to be one of the causes. Studies reveal functional abnormalities in the cingulate and basal ganglia.

Radiological findings suggest that GTS involve a decreased volume of putamen and globus pallidus.

The major neuropsychological abnormalities that have been found in the areas of attention, memory, language and motor skills. Executive functions are suspected to be affected with GTS.

Cerebellar Syndrome

There are two distinguishable cerebellar syndromes—midline and hemispheric.

Midline syndromes are characterized by imbalance. Persons are unsteady they are unable to stand with eyes open or closed and have gait disturbances. Severe midline disturbance causes "trunk ataxia" a syndrome where a person is unable to sit on their bed without steadying himself or herself. Midline cerebellar disturbances also often affect eye movements. There may be nystagmus ocular dysmetria and poor pursuit.

Hemispheric cerebellar syndromes are characterized by incoordination of the limbs. There may be decomposition of movement, dysmetria and rebound. Dysdiachokinesis is the irregular performance of rapid alternating movements. Intention tremors may be present on an attempt to touch an object. A kinetic tremor may be present in motion. Speech may be dysarthric or have irregular emphasis on syllables.

Thalamic Syndrome

Thalamic syndrome (dejerine-roussy) is a rare neurological disorder that occurs as a result of damage to the thalamus a part of the brain that affects sensation. Primary symptoms are pain and loss of sensation usually in the face arm or leg.

REFERENCES

- [1] Carpenter, M.B. (1976). Human neuroanatomy (7/e). USA: The Williams and Wilkins company
- [2] Grant, M.D., & Adams, K.M. (1996). Neurological assessment of neuropsychiatric disorder. New York: Oxford University Press.
- [3] Joseph, R. (1990). Neuropsychology, neuropsychiatry, and behavioral neurology. New York: Plenum Press.
- [4] Kandel, E.R., Schwartz, J.H., & Hassel, T.M. (2000). Principles of neural science (4/e). U.S.A: McGraw-Hill Company.
- [5] Snell, R.S. (2001). Clinical neuroanatomy for medical students (5/e). U.S.A: Lippincott, Williams and Wilkins.