

DEGENERATIVE DISEASES OF THE
CENTRAL NERVOUS SYSTEM

THESIS

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Submitted by

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Introduction and Aim of the work

In the nervous system there exists a relatively large number of lesions which consist in more or less selective degenerative phenomena unassociated with any histological indication of their etiology. In some of them there is a definite hereditary or familial basis. But the lesions may not make their appearance for twenty years or more after birth and indeed in some cases they only become apparent with the onset of senility.

Various investigators have considered them due to toxic process or due to an error in cellular metabolism.

With the exception of the senile degenerative process none of the lesions is commonly seen.

The aim of this work is to review the literature regarding classification, etiology, pathogenesis and pathology of the different degenerative diseases of the central nervous system.

ANATOMY

According to Carpenter M.B. (1976) the central nervous system is represented by the brain and spinal cord.

The Brain

It is the most complex and largest mass of nervous tissue in the body and contains billions of nerve cells. The major subdivision of the brain are fore brain, mid brain and hind brain.

Fore brain : This includes the cerebrum, thalamus and Hypothalamus.

The cerebrum is the largest portion of the brain. represents approximately seven-eighths of its total weight. nerve centers governing all sensory, motor activities, as well as poorly defined areas which determine reason, memory, and intelligence are located in the cerebrum. There are two cerebral hemispheres. These hemispheres are mirror twins. Each with a full set of

centers for sensory and motor activities of the body. Fiber tracts connecting the two hemispheres are called commissural fiber tracts. The corpus callosum is the largest of the commissural tracts. Association fiber tracts connect parts of the same hemisphere. Connecting the cortex with other parts of the brain and spinal cord are projection fiber tracts. Each hemisphere is divided into different lobes :

The parietal lobe : This lobe extends from the central sulcus about four-fifths of the way around the cortex and joins the occipital lobe at the posterior aspect of the brain.

The temporal lobe : This lobe lies beneath the lateral sulcus. It includes area 41, the sensory receptive area for auditory impulses and area 41,42, the correlative centers for auditory impulses.

The occipital lobe : Occupies the posterior segment of the cerebral hemisphere. Actually no true separation exists between the occipital lobe and the parietal and temporal lobes, although the parieto - occipital sulcus is considered the anterior margin. Area 17, the primary visual area, and adjacent area 18 and 19, the correlation centers for visual impulses, are found in

the occipital lobe.

The basal ganglia : The basal ganglia are subcortical nuclear masses derived from the cerebrum. Structures composing the basal ganglia are the caudate nucleus, the putamen, the globus pallidus and the amygdaloid nuclear complex. The caudate nucleus, putamen and globus pallidus constitute the corpus striatum. The term lentiform nucleus refers to putamen and the globus pallidus. The lentiform nucleus, with the size and shape of a Brazil nut, in transverse sections appears as a wedge with the apex directed medially. This nuclear mass lies between the internal and external capsules. A slightly curved vertical lamina of white matter divides the lentiform nucleus into an outer portion, the putamen and an inner portion, the globus pallidus.

The thalamus : is a paired structure, consists of large masses of gray matter located below the corpus callosum joined in the mid line.

The hypothalamus : lies beneath the thalamus

Mid-brain : It is found between the fore brain and hind-brain. Several nuclear masses are located on its posterior surface, the tectum, above the

cerebral aqueduct. Four of these nuclear masses are present as small elevations: The upper two, or superior colliculi, are involved in visual reflexes, especially the coordination of tracking movements, and the lower two, or inferior colliculi are associated with hearing. The four are collectively known as the corpora quadrigemina.

Two large diverging stalks emerging ventrally from each half of the cerebrum form the anterior part of the mid-brain.

The stalks are called cerebral peduncles. They constitute the main motor connection between the fore brain and the hind brain. The mid portion of the mesencephalon, known as the tegmentum, contains the red nucleus, which is connected with the cerebellum.

Hind-brain : Includes the cerebellum pons and medulla oblongata.

The cerebellum: Occupies the posterior cranial fossa. It is separated from the cerebral hemispheres by the tentorium cerebelli. The cerebellum is oval in shape with a central constriction and lateral expanded portions. The constricted central portion is

called the vermis and the lateral expanded portions, the hemispheres. The cerebellum resembles the cerebrum in the structure, with the gray matter forming a layer of cortex placed on the surface rather than centrally located, as in the spinal cord. The cerebellum is divided into lobes by deep and distant fissures. These lobes include the anterior, posterior, flocculonodular lobes.

The pons : Lies anterior to the cerebellum and between the mid-brain and medulla. On its ventral surface is a mid line groove for the basilar artery. The pons is a bridge like structure, consisting almost entirely of white matter linking the various parts of the brain.

The medulla oblongata : The most caudal basic subdivision of the brain stem, extends from the level of the foramen magnum to the caudal border of the pons. The transition from spinal cord to medulla is gradual and characterised by (1) obliteration of the anterior median fissure ventrally and the decussation of the medullary pyramids, (2) the appearance of the gracilis and cuneate tubercles dorsally, (3) the disappearance of cranial nerves and (4) the development of the fourth ventricle.

Cranial nerves associated with the medulla are:
(1) hypoglossal whose fibers emerge ventrally between the pyramid and the inferior olivary complex, (2) the accessory, the vagus and the glossopharyngeal whose fibers emerge from the post olivary sulcus and (3) the vestibulocochlear nerve whose separate components enter the brain stem at the junction of the pons and medulla. Auditory fibres are most dorsal and caudal and partially arch over the lateral aspect of the inferior cerebellar peduncle.

Ventricles of the brain

The lateral ventricles : The ependymal-lined cavities of the cerebral hemisphere constitute the lateral ventricles. They are arched-shaped and contain cerebrospinal fluid. Each lateral ventricle communicates with the slit-shaped, mid line third ventricle by two short channels known as the interventricular foramina.

The fourth ventricle : is a broad shallow rhomboid shaped cavity overlying the pons and medulla that extends from the central canal of the upper cervical spinal cord to the cerebral aqueduct of the mid-brain. Its roof is the cerebellum and the supe-

rior and inferior medullary veli, which extend toward an apex within the cerebellum known as the fastigium. The rhomboid fossa which forms the floor of the fourth ventricle is divided by the median sulcus limitans divides each half into a medial eminence and a lateral region known as the vestibular area.

The third ventricle : is a cleft found between the two thalami. It extends from the lamina terminalis anteriorly to the pineal body posteriorly . It is communicated with the lateral ventricle by the inter-ventricular foramen and with the fourth ventricle by the aqueduct. Through both communications the ependymal lining of the three ventricles becomes continuous.

Brain stem

On the anterior surface of the brain stem the medulla, pons, mid brain and part of the hypothalamus can be identified. The thalamus and epithalamus are seen in the posterior view of the brain stem. Along the dorsomedial margin of the thalamus is the sceria medullaris. The tegmentum of the mid - brain, pons, medulla represents the brain stem reticular formation. Both the reticular formation and the cort-

ically derived ventral fiber system are continuous within the brain stem, but undergo change and modification at various levels.

The meninges : The brain is invested by various membranes, floated in a clearfluid and encased in a bony vault. Three membranes surround the brain . The most external is a dense connective tissue envelope known as the dura matter or packymeninx . The innermost connective tissue membrane is the pia matter, a thin translucent membrane, adherent to the surface of the brain and spinal cord, which accurately follows every contour. Between these membranes is a delicate layer of reticular fibers forming a web like membrane, the arachnoid. The pia matter and arachnoid have a similar structure and collectively are called the leptomeninges.

The Spinal Cord

The spinal cord is a long, cylindrical structure, invested by meninges, which lies in the vertebral canal. It extends from the foramen magnum , where it is continuous with the medulla, to the lower border of the first lumbar vertebrae. The spinal cord has two enlargements, cervical and

lumbar, each associated with nerve roots which innervate, respectively, the upper and lower extremities. Caudal to the lumbar enlargement, the spinal cord has a conical termination, the conus medullaris. A condensation of pia matter, extending caudally from the conus medullaris, forms the filum terminal, the latter structure penetrates the dural tube at the levels of the second sacral vertebra, becomes invested by dura and continues as the coccygeal ligament to the posterior surface of the coccyx. The spinal cord is considered to consist of 31 segments 8 cervical, 12 thoracic, 5 lumbar, 5 sacral and 10 coccygeal.