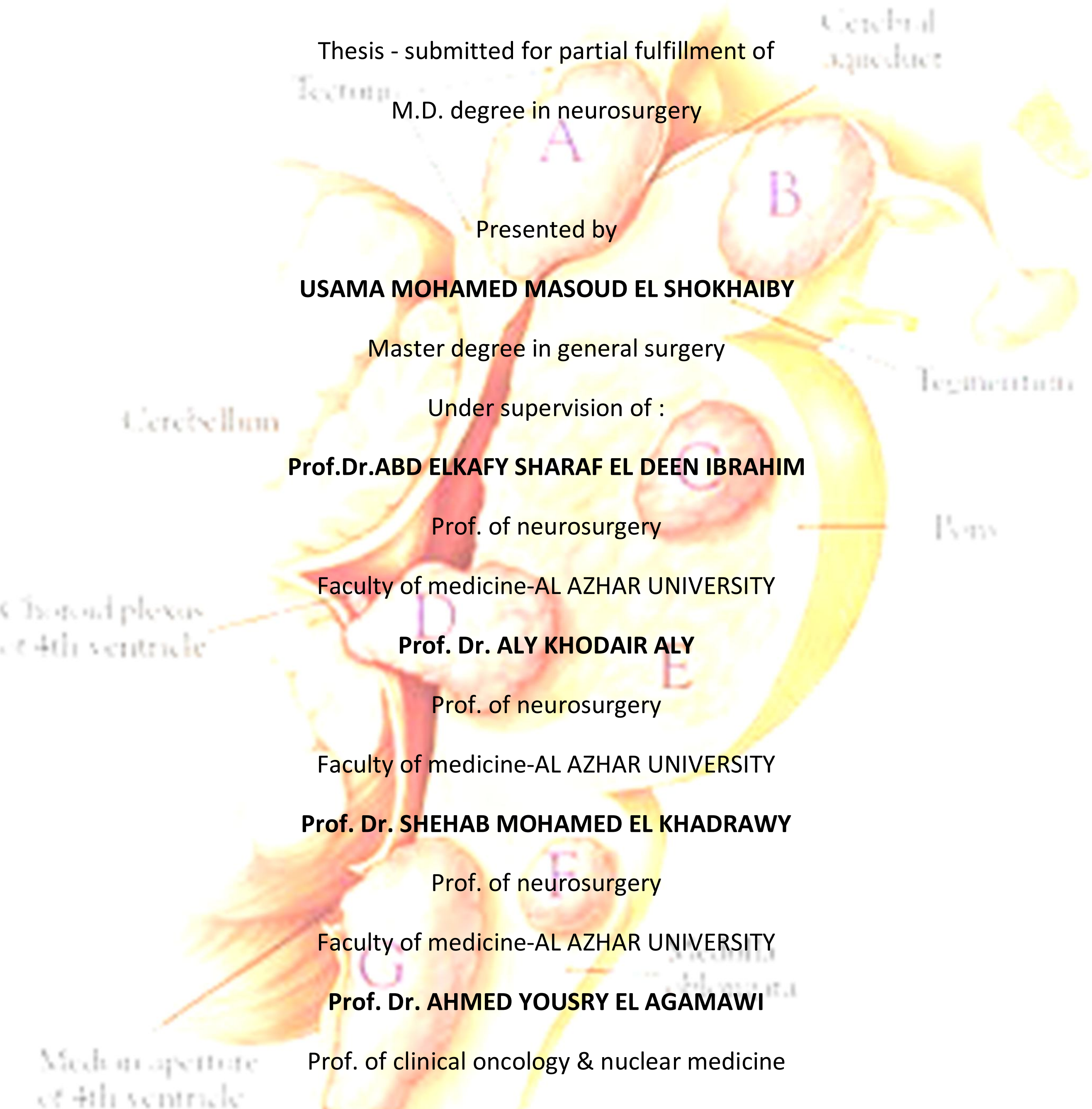
MANAGEMENT OF BRAINSTEM GLIOMAS



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M.D. degree in neurosurgery

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DEDICATION

TO MY MOTHER

AND

TO THE SOUL OF MY FATHER

ACKNOWLEDGEMENT

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To all those I say:

جزاكم الله خيرا

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INTRODUCTION

The brainstem, or mesencephalon, is defined as the midbrain, pons, and medulla.*(Gallo etal 2004)*"Brainstem glioma" is the most frequent tumor of the region but it constitutes a heterogeneous group of tumors with variable prognoses.*(Laigle-Donadey et al 2011)* Brainstem gliomas affect all ages, although they are predominant in children. There are several grading systems but they are often divided in four subgroups: tectal, diffuse pontine, cervicomedullary and dorsal exophytic. While diffuse pontine gliomas usually are high-grade tumors, the other three subtypes are usually low- grade lesions (pilocytic astrocytoma or grade II gliomas). *(Lima 2011)*

Gliomas within the brainstem constitute **10–20%** of all ***pediatric*** CNS tumors*(Gallo et al 2004)*.There are few studies of brainstem gliomas in ***adults*** since they are a rare entity, representing less than **2%**of gliomas in this population*(Guillamo et al 2001)*. In the pre-imaging era, they were considered a single entity and inoperable, but newer imaging techniques allowed the classification of subgroups with distinct behaviors. Nowadays, the knowledge of course of the disease as well the outcome of these different subtypes permits a better determination of which patients would benefit from surgery. In addition, despite the involvement of a crucial area of the central nervous system, modern neurosurgical procedures currently permit safer resections with minimal additional morbidity (*Lima 2011*).

Many classification schemes have been described for brainstem tumors. The earliest classifications relied on ***computed tomography*** and surgical observations. Current classification schemes include ***MRI sequences***. All these systems categorize the tumor by epicenter (diffuse or focal) or imaging characteristics. The simplest classification divides these tumors into two groups, either focal or diffuse regardless of tumor epicenter. The more complex schemes subdivide these tumors by location, growth pattern ('focality'), presence of hydrocephalus or hemorrhage, and growth pattern. *(Gallo et al 2003)*

A simple classification would divide brainstem gliomas in four major categories: diffuse intrinsic gliomas, focal tectal, cervicomedullary and dorsal exophytic tumors *(Laigle-Donadey et al 2008).*

In addition to ***symptomatic*** and ***general supportive*** measures, the main ways of treatment are ***neurosurgical***, ***radiotherapy*** and ***chemotherapeutic*** measures. Resection of intrinsic diffuse brainstem tumors remains impossible despite technical advances. Biopsy also is rarely indicated. However, some authors consider that guided ***stereotactic biopsy*** is relatively "safe" and believe that an accurate diagnosis should be obtained in all cases and not only when atypical clinical or imaging features suggesting a non-neoplastic lesion. A ***stereotaxic approach*** can be a rapid and safe method for evacuation of the contents of cysts and ***Ommaya*** reservoir after reaccumulation of the cyst can also be performed. Patients with hydrocephalus may require ***ventriculostomy*** or ***ventriculoperitoneal shunting*** for symptomatic relief. Focal ***radiotherapy*** is the standard treatment for ***adult*** brainstem gliomas and can improve or stabilize patients for years. *(Laigle-Donadey et al 2011)**Chemotherapy*** comes as another option especially in pediatric low grade cases where it comes first. *(Ronghe et al 2010).*

The outcome of brainstem gliomas is related to the location and histological grade of the tumors. ***Tectal lesions*** portray a good prognosis and tumors may remain stable for years. The prognosis of ***diffuse pontine gliomas*** remains dismal despite many experimental protocols of radiation and chemotherapy.

Tumor control can be achieved in most patients with ***cervicomedullary gliomas***. ***Dorsally exophytic tumors*** have a good outcome after surgical resection. *(Lima 2011)*

AIM OF THE WORK

This study aims at assessing the cases of brain stem gliomas and their diagnosis by using all the available investigations and their management according to different modalities of treatment in a trial to achieve the best results for such cases.

ANATOMY

The **brain stem**, or **encephalic trunk**, is subdivided into three sections: the *medulla oblongata*(elongated spinal cord) ,the *pons* (bridge), and the *mesencephalon*(midbrain). It is this part of the brain that is underlain by the chorda dorsalis (notochord) during embryonic development and from where ten pairs of genuine peripheral nerves (cranial nerves III – XII) emerge. (Kahle & Frotscher 2003)

The brainstem has three broad functions: (Kahle & Frotscher 2003) it serves as a conduit for the ascending tracts and descending tracts connecting the spinal cord to the different parts of the higher centers in the forebrain; (Snell 2010) it contains important reflex centers associated with the control of respiration and the cardiovascular system and with the control of consciousness; and (Naidich et al 2009) it contains the important nuclei of cranial nerves III through XII. (Snell 2010)

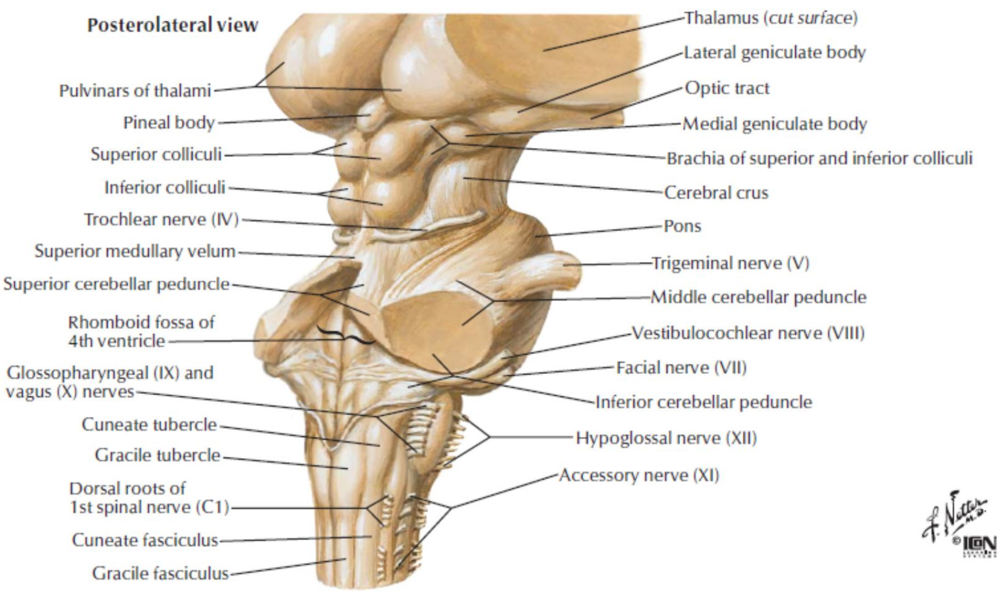

Fig.1: Brainstem (posterolateral view) (Netter et al 2002).

Gross Appearance Of the Medulla Oblongata:

The medulla oblongata connects the pons superiorly with the spinal cord inferiorly. The junction of the medulla and spinal cord is at the origin of the anterior and posterior roots of the first cervical spinal nerve, which corresponds approximately to the level of the foramen magnum. The medulla oblongata is conical in shape, its broad extremity being directed superiorly .The central canal of the spinal cord continues upward into the lower half of the medulla; in the upper half of the medulla, it expands as the cavity of the fourth ventricle.

On the anterior surface of the medulla is the anterior median fissure, which is continuous inferiorly with the anterior median fissure of the spinal cord . On each side of the median fissure, there is a swelling called the pyramid. The pyramids are composed of bundles of nerve fibers, called corticospinal fibers, which originate in large nerve cells in the precentral gyrus of the cerebral cortex. The pyramids taper inferiorly, and it is here that the majority of the descending fibers cross over to the opposite side, forming the decussation of the pyramids (Fig.2). The anterior external arcuate fibers are a few nerve fibers that emerge from the anterior median fissure above the decussation and pass laterally over the surface of the medulla oblongata to enter the cerebellum. Posterolateral to the pyramids are the olives, which are oval elevations produced by the underlying inferior olivary nuclei. In the groove between the pyramid and the olive emerge the rootlets of the hypoglossal nerve. Posterior to the olives are the inferior cerebellar peduncles (Fig.1), which connect the medulla to the cerebellum. In the groove between the olive and the inferior cerebellar peduncle emerge the roots of the glossopharyngeal and vagus nerves and the cranial roots of the accessory nerve (Fig.1).The posterior surface of the superior half of the medulla oblongata forms the lower part of the floor of the fourth ventricle . The posterior surface of the inferior half of the medulla is continuous with the posterior aspect of the spinal cord and possesses a posterior median sulcus. On each side of the median sulcus, there is an elongated swelling, the gracile tubercle, produced by the underlying gracile nucleus .Lateral to the gracile tubercle is a similar swelling, the cuneate tubercle, produced by the underlying cuneate nucleus(Fig.1) .(Snell 2010)

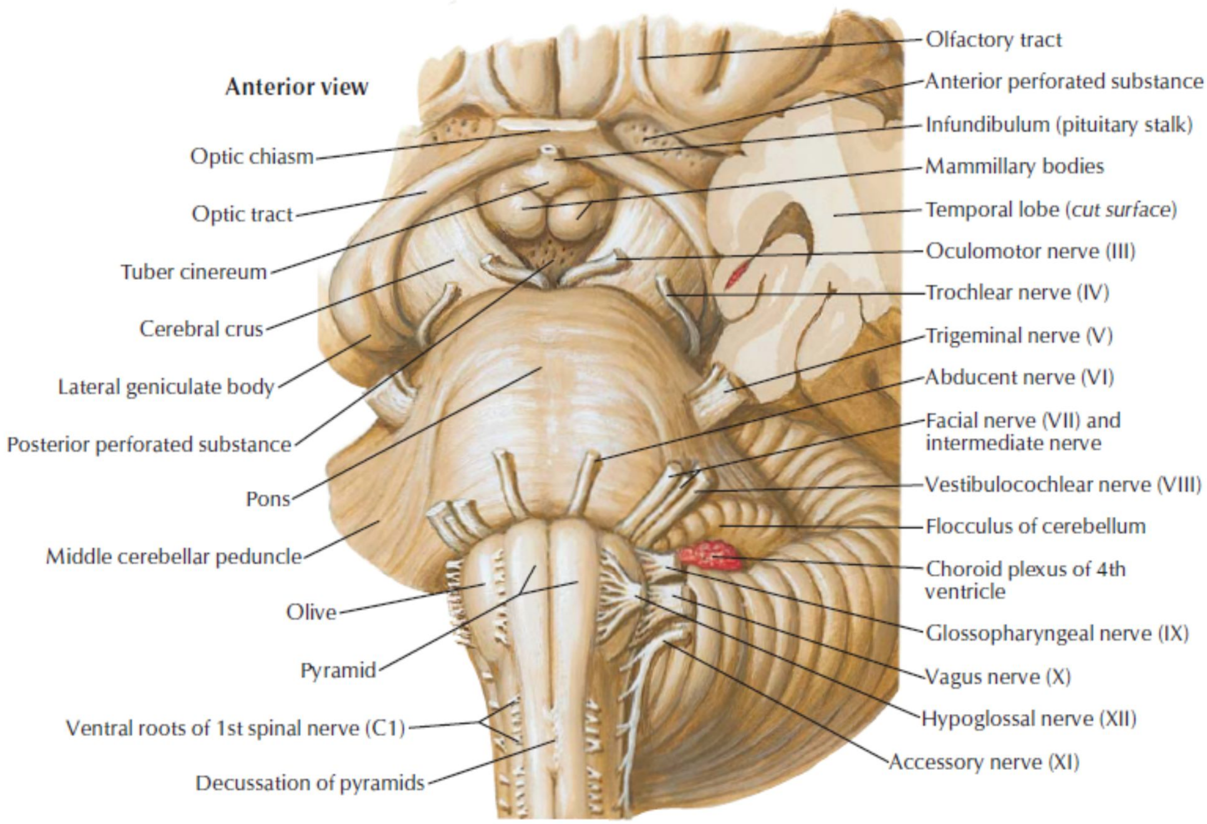

Fig.2: Brainstem(Anterior view) (Netter et al 2002)

Internal Structure Of Medulla Oblongata:

Table (1): The Different Levels of the Medulla Oblongata Showing the Major Structures at Each Level[a] (Snell 2010)

Level	Cavity	Nuclei	Motor Tracts	Sensory Tracts
Decussation of pyramids	Central canal	Nucleus gracilis, nucleus cuneatus, spinal nucleus of cranial nerve V, accessory nucleus	Decussation of corticospinal tracts, pyramids	Spinal tract of cranial nerve V, posterior spinocerebellar tract, lateral spinothalamic tract, anterior spinocerebellar tract
Decussation of medial lemnisci	Central canal	Nucleus gracilis, nucleus cuneatus, spinal nucleus of cranial nerve V, accessory nucleus, hypoglossal nucleus	Pyramids	Decussation of medial lemnisci, fasciculus gracilis, fasciculus cuneatus, spinal tract of cranial nerve V, posterior spinocerebellar tract, lateral spinothalamic tract, anterior spinocerebellar tract
Olives, inferior cerebellar peduncle	Fourth ventricle	Inferior olivary nucleus, spinal nucleus of cranial nerve V, vestibular nucleus, glossopharyngeal nucleus, vagal nucleus, hypoglossal nucleus, nucleus ambiguus, nucleus of tractus solitarius	Pyramids	Medial longitudinal fasciculus, tectospinal tract, medial lemniscus, spinal tract of cranial nerve V, lateral spinothalamic tract, anterior spinocerebellar tract
Just inferior to pons	Fourth ventricle	Lateral vestibular nucleus, cochlear nuclei		No major changes in distribution of gray and white matter

[a]Note that the reticular formation is present at all levels.

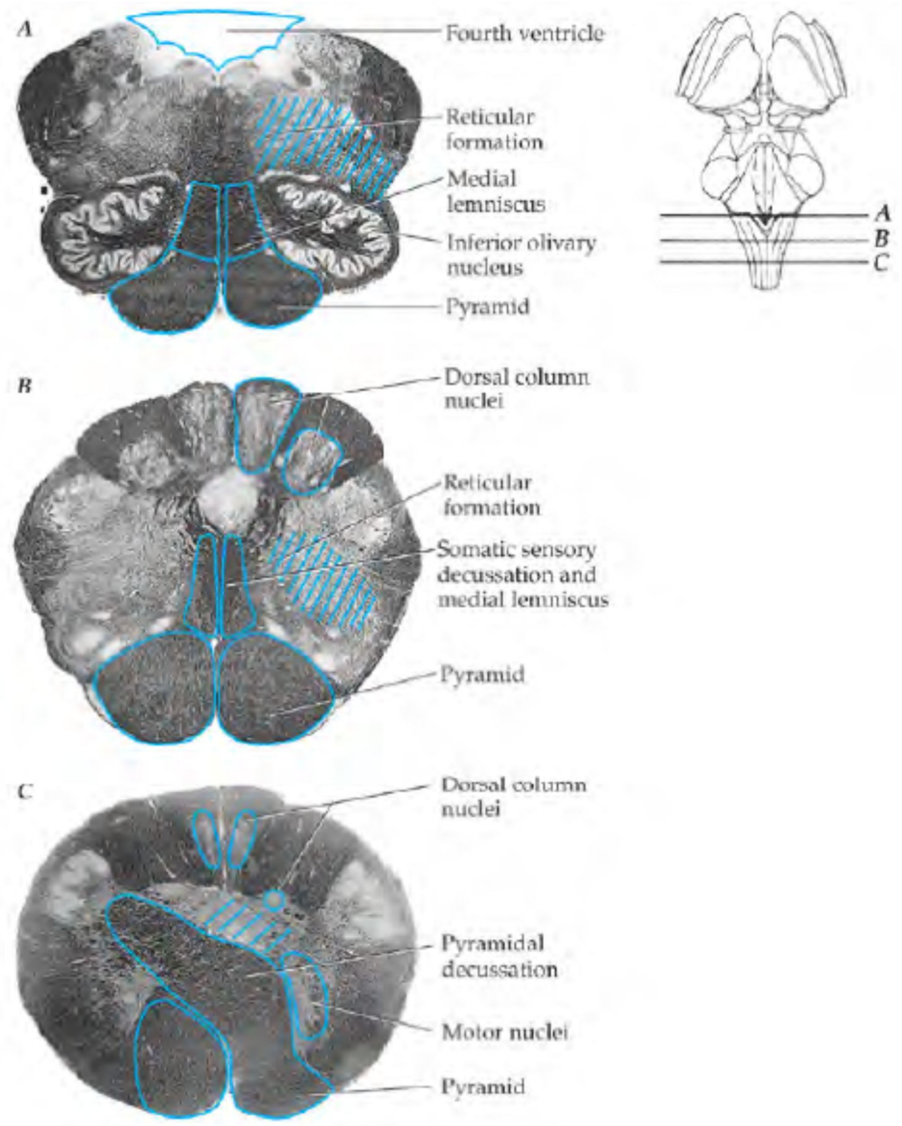

Fig.(3):Myelin stained sections through three levels of the medulla. From rostral to caudal through the ;inferior olivary nucleus(A) ,dorsal column nuclei(B),and pyramidal decussation(C).planes of the sections are indicated in the inset.(Martin 2003).

Gross Appearance of the Pons:

The pons is anterior to the cerebellum (Fig.2) and connects the medulla oblongata to the midbrain. It is about 1 inch (2.5 cm) long and owes its name to the appearance presented on the anterior surface, which is that of a bridge connecting the right and left cerebellar hemispheres.

The anterior surface is convex from side to side and shows transverse fibers that converge on each side to form the middle cerebellar peduncle (Fig.2). There is a shallow groove in the midline, the basilar groove, which lodges the basilar artery. On the anterolateral surface of the pons, the trigeminal nerve emerges on each side. Each nerve consists of a smaller, medial part, known as the motor root, and a larger, lateral part, known as the sensory root. In the groove between the pons and the medulla oblongata, there emerge, from medial to lateral, the abducent, facial, and vestibulocochlear nerves (Fig.4). (Snell 2010)

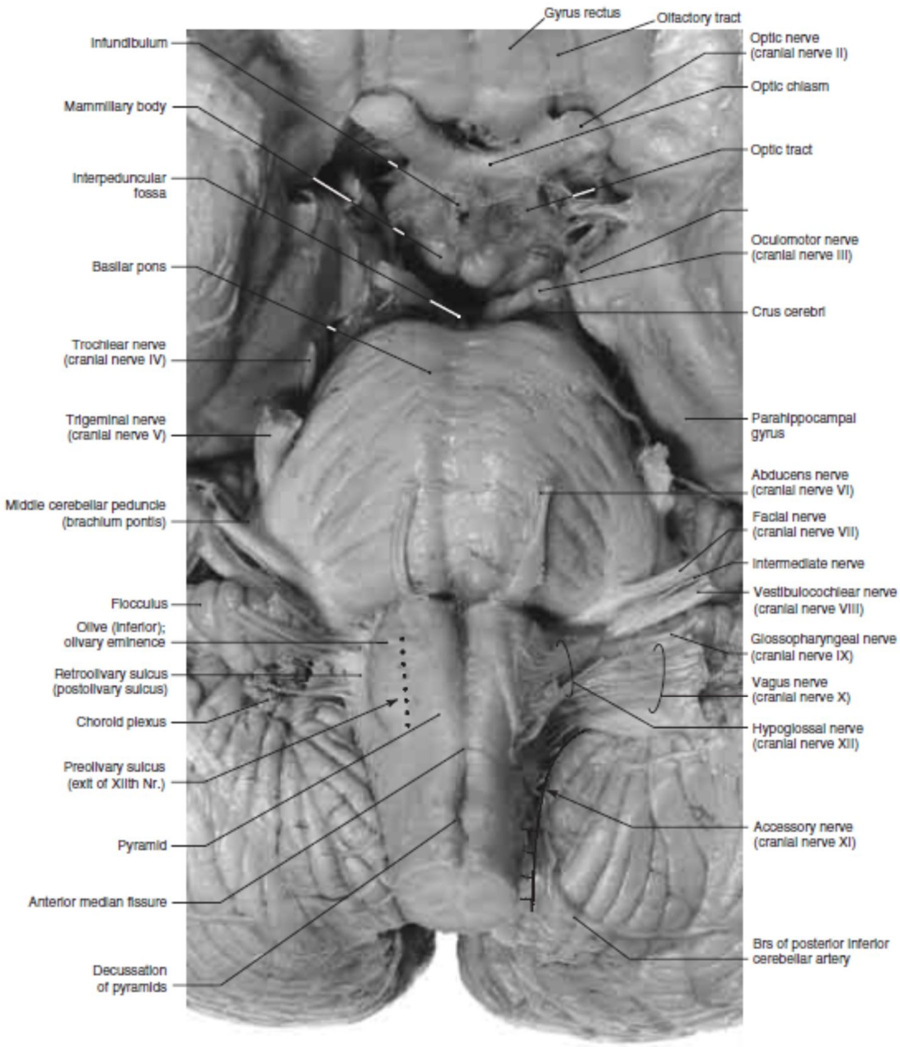

Fig.4: Detailed ventral view of the diencephalon and brainstem with particular emphasis on cranial nerves and related structures. The dots on the left side represent the approximate position of the roots of the hypoglossal nerve on that side; the general position of the (spinal) accessory nerve is shown on the right by the dark line.(Haines 2003)

The posterior surface of the pons is hidden from view by the cerebellum (Fig.5) It forms the upper half of the floor of the fourth ventricle and is triangular in shape. The posterior surface is limited laterally by the superior cerebellar peduncles and is divided into symmetrical halves by a median sulcus. Lateral to this sulcus is an elongated elevation, the medial eminence, which is bounded laterally by a sulcus, the sulcus limitans (Fig.5) .The inferior end of the medial eminence is slightly expanded to form the facial colliculus, which is produced by the root of the facial nerve winding around the nucleus of the abducent nerve (Fig.6). The floor of the superior part of the sulcus limitans is bluish-gray in color and is called the substantia ferruginea; it owes its color to a group of deeply pigmented nerve cells. Lateral to the sulcus limitans is the area vestibuli produced by the underlying vestibular nuclei (Fig.5). (Snell 2010)

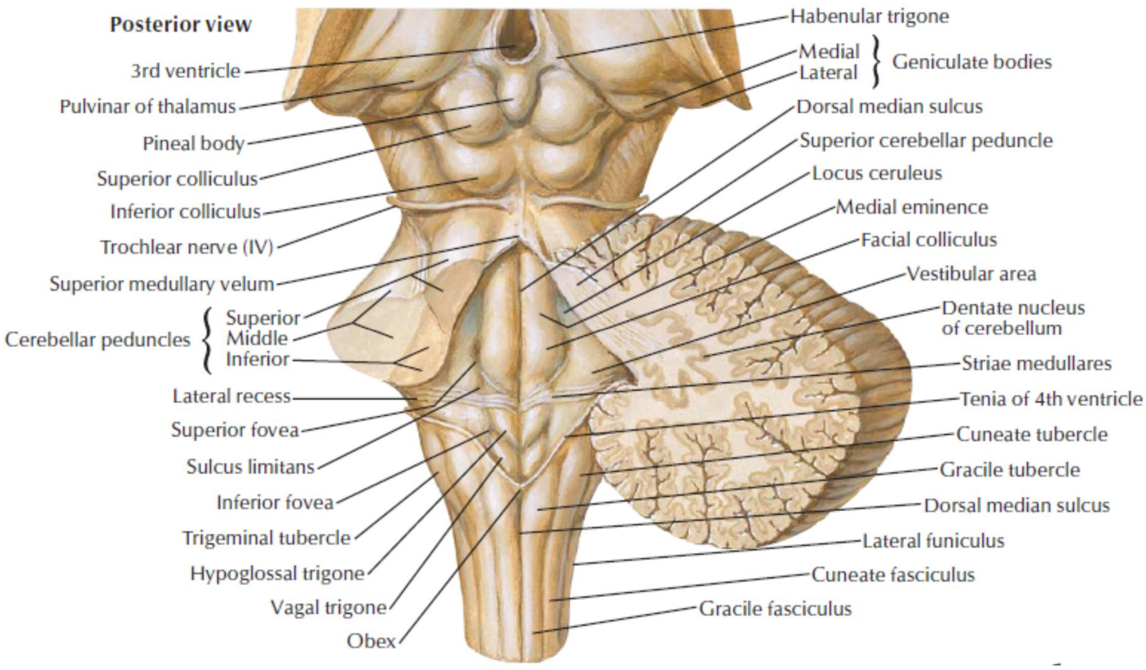

Fig.5: Brainstem (posterior view)(Netter et al 2002).

Internal Structure of the Pons:

Table2: The Different Levels of the Pons Showing the Major Structures at Each Level[a] (Snell 2010)

Level	Cavity	Nuclei	Motor Tracts	Sensory Tracts
Facial colliculus	Fourth ventricle	Facial nucleus, abducent nucleus, medial vestibular nucleus, spinal nucleus of cranial nerve V, pontine nuclei, trapezoid nuclei	Corticospinal and corticonuclear tracts, transverse pontine fibers, medial longitudinal fasciculus	Spinal tract of cranial nerve V; lateral, spinal, and medial lemnisci
Trigeminal nuclei	Fourth ventricle	Main sensory and motor nucleus of cranial nerve V, pontine nuclei, trapezoid nuclei	Corticospinal and corticonuclear tracts, transverse pontine fibers, medial longitudinal fasciculus	Lateral, spinal, and medial lemnisci

[a]Note that the reticular formation is present at all levels.

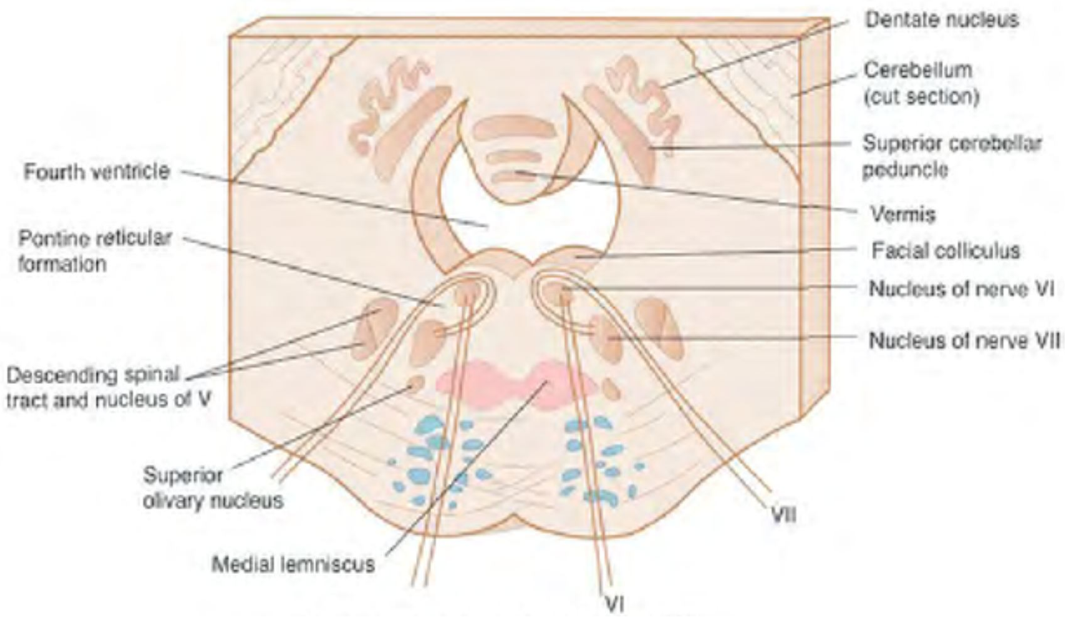

Fig.(6):Lower pons;level of 6th and 7th nerves.(Waxman 2010)