Midline Skull Base Tumors

Essay

Submitted for partial fulfillment of master degree

In

Otorhinolaryngology

By

Amr Mohamed Hassan

(M.B.B.Ch, Cairo University)

Under supervision of

Prof. Reda Hussein Kamel

Professor of Otorhinolaryngology

Cairo University

Prof. Al Gohary Mohamed Al Gohary

Professor of Neurosurgey

Cairo University

Dr. Mahmoud Attia Mohamed

Assistant professor of Otorhinolaryngology

Cairo University

2008

Dedication

To
My parents
And
My brother Hesham

Acknowledgments

Thanks to **God**

I would like to express my sincere thanks and supreme gratitude to **Prof. Reda Hussien Kamel,** professor of otorhinolaryngology, Cairo University for suggesting the subject of the essay and for his valuable time and advices he offered me through the preparation of this work.

My appreciations and cordial thanks are offered to **Prof. Al Gohary Mohamed Al Gohary,** professor of neurosurgery, Cairo University for his supervision and constructive criticism of all vital steps of this work.

I am also too much grateful to **Dr. Mahmoud Attia Mohamed,** assistant professor of otorhinolaryngology, Cairo University for his active participation and guidance throughout the preparation of the work

I am also greately indebted to all my family members for their active support and encouragement

****

ABSTRACT

Conventional fractionated **radiotherapy** has been used primarily for malignant skull base tumors and radiosensitive tumors of the intermediate malignant type. Radiation therapy also reduces the growth and recurrence rates of some of the benign tumors including paragangliomas. There are three basic forms of stereotactic radiosurgery represented by three different technological instruments. Each instrument operates differently, has a different source of radiation and may be more effective under different circumstances. The three are: A) Particle beam (proton), B) Cobalt-60 based (photon) and C) Linear accelerator based (linac).

Chemotherapy is indicated in specific malignant tumors including nasopharyngeal carcinomas, melanomas and rhabdomyosarcomas.

Hormonal therapy for meningiomas and juvenile angiofibromas, which often carry a hormonal receptor, is still in the investigative stage**.** Interferon, somatostatin analogues, monoclonal antibodies, and gene therapies are likewise considered experimental.

To obtain the maximum effect of treatments, various **combined treatment** regimens are indicated for the management of certain skull base tumors.

Key Word:

Midline Skull Base Tumors
Chemotherapy
Hormonal therapy
radiotherapy

Contents

List of figures

Figure Number: Title

List of figures

Figure Number: Title

List of figures

Figure Number: Title

List of abbreviations

- ABR ------------------- Auditory Brainstem response
- ACC------------------- Anterior Skull Base
- CG--------------------- Cholesterol Granuloma
- CN ---------------------Cranial Nerve
- CSF--------------------.Cerebro-Spinal Fluid
- CT----------------------Computed Tomography
- EMA--------------------.Epithelial Membrane Antigen
- GSPN------------------Greater Superficial Petrosal Nerve
- HRCT-------------------High Resolution Computed Tomography
- IAC---------------------Internal Auditory Canal
- ICA---------------------Internal Carotid Artery
- IOF--------------------- Inferior Orbital Fissure
- JNA---------------------Juvenile Nasopharyngeal Angiofibroma
- LINAC-based SRT ----Linear Accelerator based Stereotactic Radio-Therapy
- LSPN--------------------Lesser Superficial Petrosal Nerve
- MRI ---------------------Magnetic Resonant Image
- ONB---------------------Olfactory Neuro-Blastoma
- PgR--------------------- Progesterone Receptors
- PLGA--------------------polymorphous low-grade adenocarcinoma
- SOF----------------------Superior Orbital Fissure
- VAC----------------------Vincristine- D-Actinomycin- Cyclophosphamide
- VA------------------------Vertebral Artery
- NHL----------------------Non-Hodgkin's lymphoma

****

Introduction

The skull base is a highly complex region with multiple bony foramina through which pass several important neurovascular structures (Cavallo et al, 2005). The skull base can be divided arbitrarily into anatomic subsites corresponding to the major intracranial compartments as follows: (1) anterior skull base (anterior cranial fossa), (2) middle skull base (middle cranial fossa), (3) posterior skull base (posterior cranial fossa), and (4) central skull base (brainstem) (Jho and Alfieri, 2002).

Because all the neurovascular connections between the brain and the neck pass through the foramina within the cranial base, skull base tumors are surrounded by multiple vital structures; therefore, safe removal poses a considerable surgical challenge (Weiss MH, 1987).

As a whole, tumors of the skull base are uncommon entities, and no accurate data regarding the incidence of skull base neoplasms are available.

The clinical presentation of skull base tumors varies greatly and relates directly to the location of the lesion and the growth rate. It ranges from local pain, headache, nasal obstruction and recurrent sinusitis to signs of increased intracranial tension, cranial nerve palsies and cerebrospinal fluid rhinorrhea (Morita et al, 2000).

Biopsy is the only sure technique that allows tissue diagnosis of skull base tumors.However, in many cases, the clinical presentation and diagnostic imaging provide sufficient information upon which to base a treatment decision (Jho and Ha, 2004).

Because surgical excision is the treatment of choice for the vast majority of benign skull base tumors, where a wide surgical margin is unnecessary, a histologic tissue diagnosis is unnecessary prior to definitive resection. However, an attempt at biopsy is indicated for cases in which diagnostic imaging has not sufficiently narrowed the diagnostic possibilities. Resection of lesions involving this area requires a variety of innovative skull base approaches. These include anterior, anterolateral, and posterolateral routes, performed either alone or in combination and resection via these routes often requires extensive neurovascular manipulation. (Cavallo et al, 2005).

Over the past 20 years, advances in anesthesia, improvements in surgical technology, and refinements in reconstructive modalities have enabled surgical removal of cranial base tumors that were previously associated with unacceptable morbidity. The constant improvements in diagnostic imaging techniques and the increasing use of image guidance systems during endoscopic endonasal procedures has provided increasing accuracy, safety and allowed an improved, constant surgical orientation in an anatomically complex area. (Cavallo et al, 2005).

Recent anatomical studies and clinical reports have detailed the anatomy encountered in the endoscopic endonasal approach (Jho and Alfieri, 2002, Alfieri et al, 2003, Liu et al, 2003).

The major potential advantage of the endoscopic approach to the skull base is that it provides a direct anatomical route to the lesion without traversing any major neurovascular structures and obviating any brain retraction. The extended endoscopic endonasal route, given its versatility, offers the possibility of exposing the entire midline skull base from below, with the advantage of passing through a less delicate structure (nasal cavity) to reach a more noble one (the brain with its neurovascular structures). Furthermore, the endoscope enables the surgeon to work on tumors located in supra, para, retro, and infrasellar regions under direct visual control (De Divitiis et al, 2002).

The potential complications or difficulties associated with the extended endonasal approach to the midline skull base are **,** increased bleeding from circular sinuses and **,** high rates of postoperative CSF leakage.

The aim of the present work is to study the different types of midline skull-base tumors and to present the recent advances in their management

****

Anatomy of skull base

Introduction

The skull base forms the floor of the cranial cavity and separates the brain from other facial structures. This anatomic region is complex and poses surgical challenges for otolaryngologists and neurosurgeons. Knowledge of the normal and variant anatomy of the skull base is essential for efficient and safe surgical treatment of disease in this area (Jho and Alfieri, 2002).

The 5 bones that make up the skull base are the ethmoid, sphenoid, occipital, frontal, and paired temporal bones. The skull base can be subdivided into 3 regions: the anterior, middle, and posterior cranial fossae (Nuss and O'Malley, 2005).

Definition of midline skull base

When viewing the skull from the inferior perspective, the central compartment may be define as that area between two parasagittal lines drawn from the medial pterygoid plate to the occipital condyle on each side. These lines correspond approximately to the pathways of the ICAs through the skull base. Thus the central compartment consists of the pituitary fossa, the sphenoid rostrum and lower sphenoid sinus, the nasopharynx, the pterygopalatine fossa and the lower portion of the clivus (see fig (1)) (Krepsi and Sisson 1984).

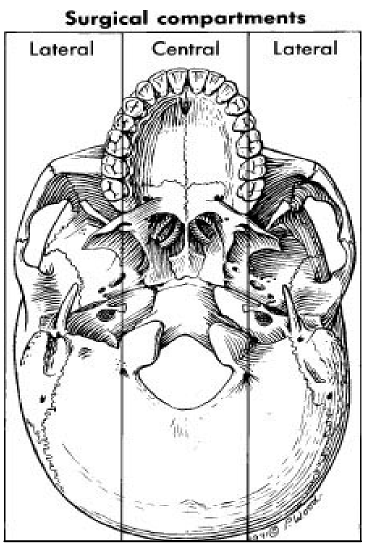

Fig (1) compartments of skull base (Krepsi and Sisson 1984).

Anterior skull base

A-Boundaries

The anterior limit of the anterior skull base is the posterior wall of the frontal sinus. The anterior clinoid processes and the planum sphenoidale, which forms part of the roof of the sphenoid sinus, mark the posterior limit. The frontal bone forms the lateral boundaries. The frontal bone houses the supraorbital foramina.

The greater portion of the anterior floor is convex and grooved by the frontal lobe gyri. This portion of the skull base consists of the orbital plates of the frontal bone. The ethmoid bone forms the central part of the floor, which is the deepest area of the anterior cranial fossa. In the center of this region is the cribriform plate, through which the olfactory nerve fibres pass. The fovea ethmoidalis, or the roof of the ethmoid cavity, continues laterally from the cribriform plate. The cribriform plate may be >1 cm lower than the roof of the ethmoid cavity (fovea ethmoidalis), and it is made of extremely thin bone compared with the relatively thick bone of the lateral fovea ethmoidalis. During transethmoidal approaches to the anterior skull base, this relationship is extremely important to remember. (Nuss and O'Malley, 2005)

The foramen caecum sits between the frontal crest and the prominent crista galli and is a site of an inconsistent communication between the draining veins of the nasal cavity and the superior sagittal sinus. The crista galli, which projects up centrally between the cerebral hemispheres, serves as the site of attachment for the falx cerebri. (Tiedemann, 1997)

The optic chiasma lies behind the planum sphenoidale. The anterior clinoid processes -the most medial part of the lesser wing of the sphenoid- shares in forming the roof of the most posterior part of the optic canal and is an important landmark for supracavernous internal carotid artery (ICA) (Goel, 1997) .

B-Inferior relationships - Extracranial aspects:

The most important anatomic structures below the anterior cranial fossa are the orbits and the paranasal sinuses.

The bony orbit is often a route for intracranial and extracranial spread of infection and tumors because of its direct proximity to the anterior fossa. The posterior wall is adjacent to the superior sagittal sinus and frontal lobe dura. The orbital apex includes the optic canal, the superior orbital fissure (SOF), and the inferior orbital fissure (IOF). The SOF conveys the oculomotor, trochlear, abducens, and ophthalmic nerves (cranial nerves [CN] III, IV, VI, and V1, respectively), as well as the ophthalmic veins. The IOF transmits the maxillary nerve (CN V2) and infraorbital vessels, and it communicates with the infratemporal and pterygomaxillary fossae. The lateral portion of the IOF is an important surgical landmark for positioning lateral orbital osteotomies during anterior skull base resections. The optic canal transmits the optic nerve (CN II) and the ophthalmic artery (Lyons, 1998).

The medial wall is closest to the apex and is formed by the orbital process of the frontal, lacrimal, ethmoid, and sphenoid bones. The medial wall transmits the anterior and posterior ethmoid arteries through their respective foramina. These foramina help in identifying the frontoethmoid suture line, which marks the inferior extent of the anterior cranial fossa. The posterior ethmoid artery foramen is also an important surgical marker for the location of the optic canal and nerve, which lies about 0.5 cm posterior to it. The greater wings of the sphenoid and the frontal process of the maxilla form the lateral walls. The posterior most segment of the lateral orbital wall forms the anterior wall of the middle cranial fossa (Mafee ,2005, Nuss and O'Malley 2005).

The ethmoid sinuses are situated inferior to the anterior cranial fossa and medial to the orbits. The frontal sinuses arise as evaginations of ethmoid air cells into the frontal bone and have a thick anterior and thinner posterior wall. The posterior wall is adjacent to the superior sagittal sinus and the frontal lobe dura. As a result, the frontal sinus can be used as a route of surgical entry into the anterior cranial fossa. Infectious processes and tumors can exploit this relationship, as well, to gain intracranial access (Netter, 1989).

C-Contents:

The dura mater attaches anteriorly at the frontal crest and crista galli to form the falx cerebri, which transmits the superior and inferior sagittal sinuses. The superior sagittal sinus drains the superior cerebral and frontal diploic veins of Breschet. These veins form a potential pathway for infection to spread intracranially, causing complications such as sagittal sinus thrombosis, empyema, or abscess. The foramen cecum, found anterior to the crista galli, usually ends blindly, though it may transmit a vein from the nasal mucosa to the superior sagittal sinus. Its patency may lead to the formation of developmental anomalies, such as nasal dermoid cysts, nasal gliomas, encephaloceles, and meningoencephaloceles (Janfaza and Nadol, 2001and Moore and Agur, 1995).

The frontal lobes occupy the anterior fossa and sit superior to the orbits and sinonasal tract. The major structures in this area are the olfactory bulb and tract. The olfactory bulb lies along the medial edge of the frontal orbital plate and connects with the olfactory tract, which courses above the cribriform plate and planum sphenoidale (see fig (2))(Janfaza and Nadol 2001).

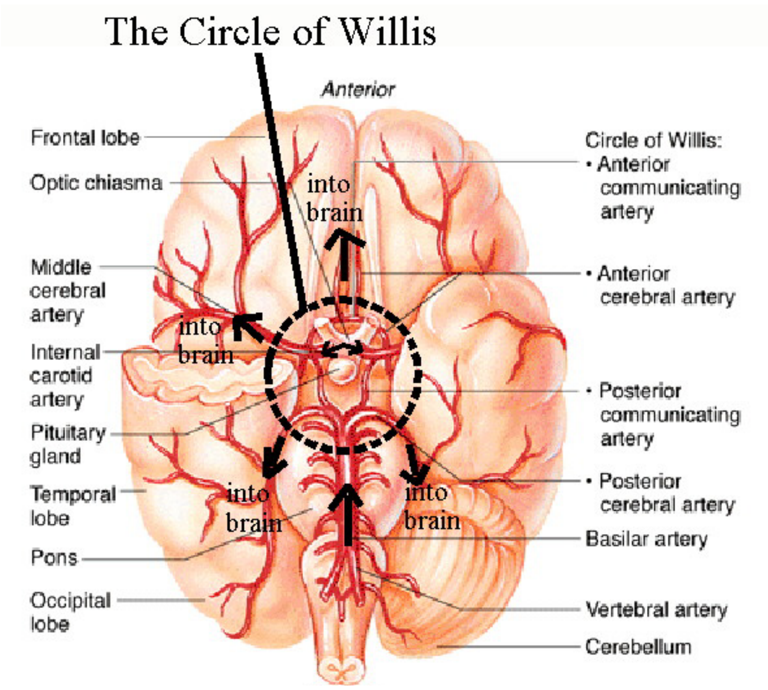

***Fig (2) ventral surface of the brain showing the circle of Willis (**Janfaza and Nadol 2001**)**

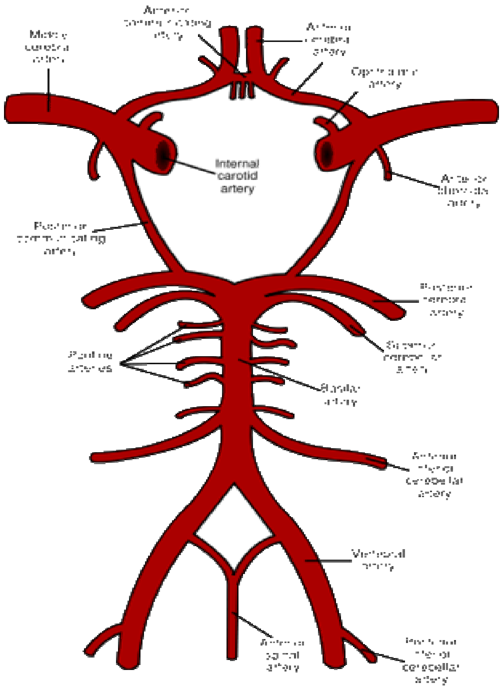

Fig (3) diagram showing the structure of the circle of Willis
(**Moore and Dalley, 1999**)

Components

- Anterior cerebral artery (left and right)
- Anterior communicating artery
- Internal carotid artery (left and right)
- Posterior cerebral artery (left and right)
- Posterior communicating artery (left and right)

The basilar artery and middle cerebral arteries, though they supply the brain, are not considered part of the circle. (See fig (3) (Moore and Dalley, 1999)

Middle skull base
1-Intracranial aspects: Boundaries

The lesser wing of the sphenoid forms the anterior limit of the middle skull base as well as the anterior clinoid process. The posterior limit is the posterior clinoid process as well as the posterior edge of the petrous bone. The greater wing of the sphenoid forms the lateral limit as it extends laterally and upward from the sphenoid body to meet the squamous portion of the temporal bone and the anteroinferior portion of the parietal bone. The greater wing of the sphenoid forms the anterior floor of the fossa. The anterior aspect of the petrous temporal bone forms the posterior floor of the middle cranial fossa (Netter, 1989 and Janfaza and Nadol 2001).

The body of the sphenoid makes up the central portion of the middle fossa and houses the sella turcica.The sella turcica can be found between the anterior and posterior clinoid processes and is composed of 3 sections. The tuberculum sellae