MANAGEMENT OF
ACOUSTIC NERVE TUMOUR

Essay Submitted for Partial
Fulfilment of the Master Degree in
Oto-Rhino-laryngology

By
Abdel Hamid Mohamed Anwar Abdel Hamid
M.B., B.Ch. (Ain Shams University)

SUPERVISED BY

Dr. Ahmad Elrefai

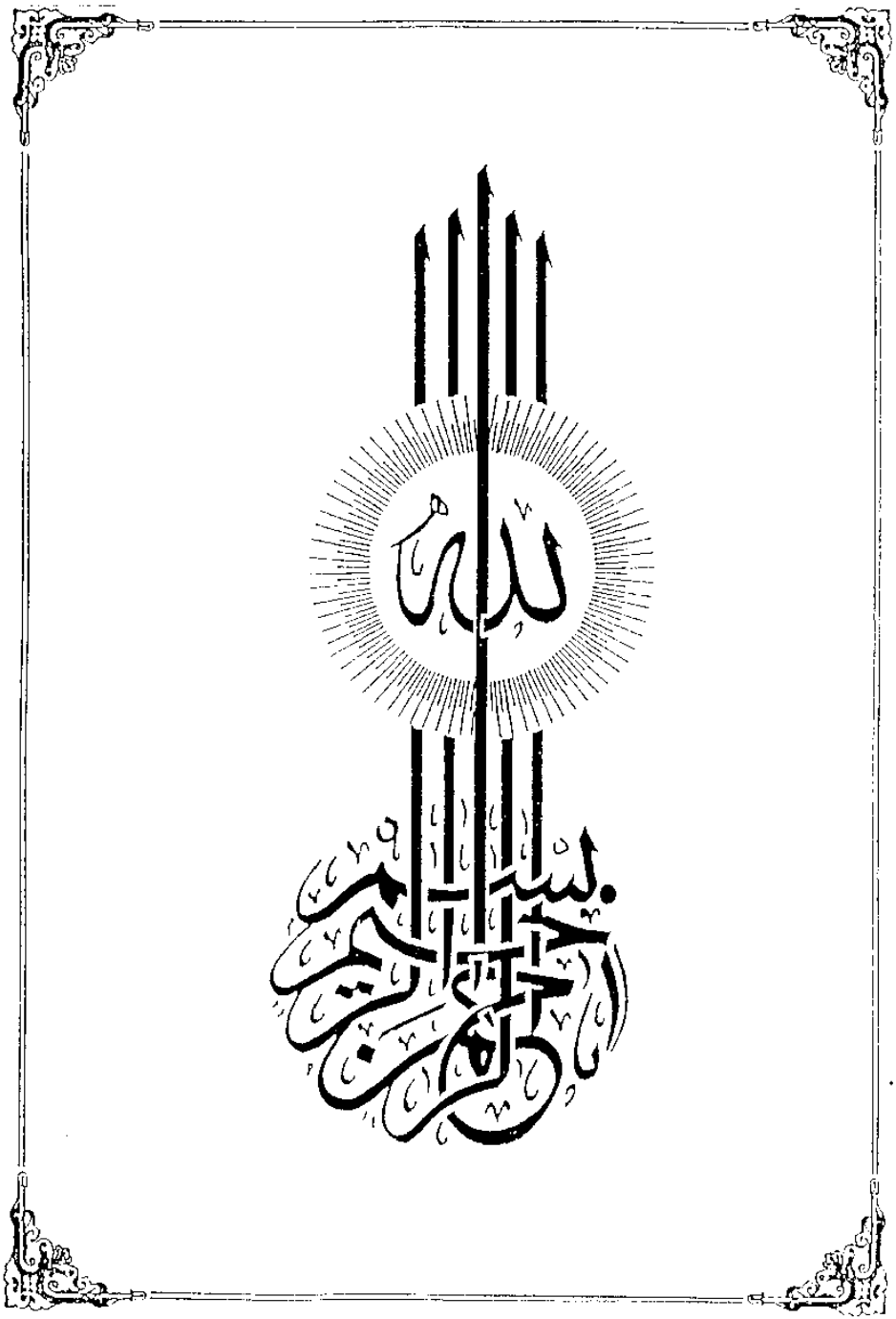
Assistant Professor of Oto-Rhino-laryngology

Faculty of Medicine
Ain Shams University
1985

This work is dedicated to every

person who learned me, to my beloved

family who supported me



ACKNOWLEDGEMENT

I would like heartly to thank and express my great gratitude to Prof. Dr. ***Ahmad Elrefai, Assistant*** *Professor of Oto-Rhino-Laryngology, Ain Shams University, for his kind help and his sincere guidance.*

I am very grateful and I would very much like to convey my true thanks to my Prof. Dr. ***Nabiel Rabie,*** *Assistant Professor of Oto-Rhino-laryngology, Ain Shams University for his help and kindness during preparation of this work.*

Finally, deep thanks to my wife who contributed me to complete this work.

The Candidate

INTRODUCTION

INTRODUCTION

Acoustic nerve tumour is a benign neoplasm which arises from Schwann sheath cells of the eighth nerve, either its vestibular or less commonly cochlear division.

It was clearly described in a patient in 1870 by Charles Bell. Acoustic nerve tumour is known as acoustic neuroma, neurinoma, neurilemmoma, perineural fibroblastoma, or better schwannoma. This confusion regarding terminology has evolved because one group of investigators believed the tumour arises from schwann cells i.e.: neuro-ectodermal in origin and another believed they are of fibroblastic mesodermal origin. However electron microscopic studies of these tumours have demonstrated that they are composed of schwann cells, and thus should be correctly called schwannoma. That schwannomas arise more commonly from the vestibular than the cochlear division of the 8th nerve, this can be explained embryologically as follows. During development the biopoler ganglion cells of the vestibuler and cochlear nerves send fibres peripherally to the auditory vesicle and centrally to the brain stem, the fibres are accompanied by neurolemmal cells but outgrow them. When

the nerve fibres reach the brain stem, glial cells grow out peripherally to meet the schwann sheath cells. The vestibuler fibres reach the brain stem earlier than the cochlear, and thus the glial cells migrate out further along the vestibuler than along the cochlear division. This is thought to result in apparent excess of schwann cells in the vestibuler nerve as compared to the cochlear nerve, and predispose the vestibuler nerve to develop schwannoma more than the cohlear nerve.

Acoustic nerve tumours represent the third largest group of intracranial tumours, after gliomas and meningiomas. They account for 8 % of all intracranial tumours and about 80 % of those occurring in the cerebello-pontine angle. Acoustic nerve tumours are most frequently encountered in the middle decades of life (35-40 years), and more commoner in females in the ratio 3:2.

CLINICAL FEATURES

CLINICAL FEATURES

Onset:

The onset of the symptoms generally occurs between the ages of 30-50 years. It tends to occur in the following phases.

A- **Otological Phase:**

The earliest and most consistent symptom of the eighth nerve tumours is :

1- **Tinitus:**

The tinitus usually develops with a loss of hearing, but it may be the only symptom (Hitselberger and House, 1971). The quality of the tinitus varies and, at times, may represent ringing bells or a roaring sound. The noise may be constant or intermittent in nature.

2- **Auditory loss:**

Frequently accompaneis the tinitus. Many patients have difficulty in discerning the meaning of sound, and distortion of hearing with the telephone is a common complaint. While hearing loss may be present for many years before patients seek medical attention, the majority of

cases have deficits present for 5 years or less (clemis and Mastricola, 1976).

In general the hearing loss is unilateral and progressive, but occasionally it may present as a sudden sensorineural hearing loss.

3- **Dizziness:**

Is a common complaint in the early stages of acoustic neuroma. It appears in the form of an unsteadiness in about 83 per cent of the patients (Pulec et al., 1964).

True vertigo is not as common; it occurs in about one third of patients, but it may become a more prominent symptom with the growth of the tumour (Hitselberger and House, 1971).

Occasionally the vertigo presents with paroxysms, as in Meniere's disease.

B- **Neurological Phase:**

The signs and symptoms depend upon the direction of enlargement taken by the tumour.

The first symptom of pontine involvement is may be:-

1- Headache: which becomes worse at night and as internal hydrocephalus increases, the headache becomes more intense.

2- Vomiting.

3- Papilloedema.

4- Other cranial nerves may be involved and gives signs and symptoms. The two cranial nerves most commonly involved are the V (fifth) and VII (seventh). The cranial nerves VI, IX, X, XI, and XII may be affected in the terminal stages.

The Vth nerve is usually the first adjacent cranial nerve to become implicated. The sensory portion is more commonly affected than the motor portion.

Sensory Portion:

a) Blunting of the corneal reflex, especially when the recumbent patient turns his head to the side of the tumour.

b) Unilateral numbness and tingling of the face.

c) Irritation may cause a form of tic doloureux.

Motor Portion:

Motor paralysis will affect the muscles of mastication with a symmetrical opening of the mouth.

The VI nerve involvement causing diplopia due to paralysis of lateral rectus muscle. The VII nerve involvement:-

A facial tic which preceeds paresis may be the presenting symptom of an acoustic neuroma but, paralysis is often a very late physical sign unless there has been a sudden rapid increase in size of an intrameatal tumour, possibly due to haemorrhage. Hitselberger and House (1966) noted hypoaesthesia of the posterior wall of the external auditory meatus in 25 cases of surgically confirmed acoustic neuromas. It is thought that this portion of the external auditory canal is supplied by sensory twigs of the facial nerve.

Facial involvement may be first noticed by a failure of blinking or a failure to bury the eye lashes on the affected side when the eye lids are closed tightly. A regulated tap on the supra-orbital region shows a delayed blinking reaction. Other aspects of facial nerve function

can be demonstrated by tests of taste and tear secretion. Electrogustometry applied to the anterior two thirds on the tongue shows a decrease in sensitivity to more than 20 U A stimulation (Pulec and House, 1964). Chemical tests of taste threshold (Hinchcliffe, 1958) are usefull but Hinchcliffe (1969) has met cases where the chemical threshold was normal and there was no response to electrical stimulation of the tongue. Other useful tests for assessing the general visceral efferent fibres of the facial nerve, which are carried by the greater superficial petrosal nerve and the chorda tympani, are the submandibular salivery flow test of Magielski and blatt (1958) and the quantitative nasolacrimol reflex measurements of Zilstorff Pederson (1959).

The IXth and Xth cranial nerves are affected later on resulting in:

* Dysphagia.
* Dysphonia.
* Inhalation of fluids.
* Palatal nystagmus, arythmical up and down movement of the soft palate may be an early indication of irritation of IXth nerve.

The XIth and XIIth cranial nerves: evidence of involvement of these nerves is uncommon.

Cerebellum: Cerebeller symptoms appear when the tumour affects the ipsilateral cerebeller lobe resulting in:

a) Incoordination of movements.

b) Nystagmus: spontaneous nystagmus is present in over 75% of cases, it is commonly horizontal, rarely vertical due to central involvement. It is first degree contralateral nystagmus, and may later become second degree accompanied by the first degree to the ipsilateral side.

c) Ataxia of the extremities on the side of the tumour.

d) Hypotonia.

e) Dysdiadokokinesis.

f) Intention tremors.

g) Staggering broad-based gait with a tendency to fall towards the side of the lesion may result from affection of the cerebllum and its connections.

Advanced tumours may be accompanied by attacks of giddiness, vomiting and jactitation followed by rigidity and unconsciousness.

c- **Terminal Phase:**

Increasing internal cranial pressure combined with bulbar palsy eventually terminate life, the patient losing control of sphincters and passing from delirium into coma with cheyne-stoke respiration.