Acoustic neuroma

A plea for early diagnosis and treatment

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Introduction

Following the report by House (1964) on the translabyrinthine approach to the internal auditory meatus, otologists have shown much interest in the early diagnosis and treatment of acoustic neuroma, a field previously considered to be the exclusive province of the neurosurgeon. Early diagnosis and prompt excision of the small tumour have led to very low operative mortality and morbidity (Pool *et al.*, 1970).

However, many surgeons still feel that acoustic neuromata are slow-growing tumours and that surgical intervention is not justified until symptoms and signs are advanced and the tumour is large. We have therefore thought it of value to review the clinical features and fate of 214 patients with large acoustic neuromata treated at the Radcliffe Infirmary, Oxford, during a thirty-year period from 1938 to 1968.

Incidence

One-hundred-and-forty-three patients were female and seventy-one were male, a ratio of 2:1. Tumours presented most commonly in the fifth decade (Fig. 1), and there was no predilection for side. Three patients had bilateral tumours and eleven patients had features of generalized neurofibromatosis.

Early symptoms

One-hundred-and-sixty-patients (75 per cent) presented with auditory symptoms (Table I), and were seen in ear, nose and throat departments. However, 25 per cent presented with neurological symptoms. In many cases there was a delay in diagnosis due to minimal initial findings, but this did not affect subsequent treatment as modern techniques for excising small tumours had not been developed. Invariably surgery was not contemplated until there was a significant neurological deficit (Pennybacker and Cairns, 1950).

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Preoperative findings

On admission for surgery all patients had symptoms and signs of a cerebellopontine angle lesion. These can be considered under six main headings (Cushing, 1917): audiovestibular; headache; cerebellar; cranial neuropathy; raised intracranial pressure; and medullary compression.

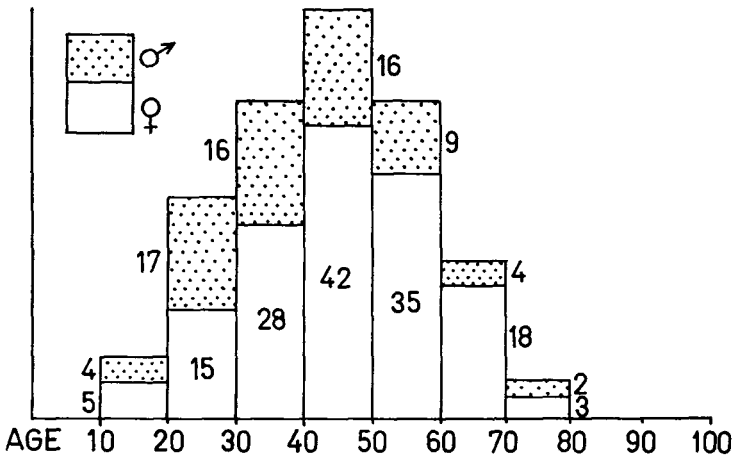


FIG. 1. Age and sex distribution in 214 patients with acoustic neuroma.

TABLE I.

PRESENTING SYMPTOMS IN 214 PATIENTS

Deafness alone	90
Deafness and tinnitus	62
Tinnitus alone	8
Total auditory symptoms	160 (75 per cent)
Headache	25
Unsteadiness	17
Failing vision	5
Facial paraesthesiae	3
Trigeminal neuralgia	1
Facial spasm	1
Convulsions	1
Loss of consciousness	1
Total neurological symptoms	54 (25 per cent)

Audiovestibular findings

Two-hundred-and-six patients had ipsilateral hearing loss, with or without tinnitus (Table II). In most cases this was gradually progressive but eleven patients suffered sudden deafness with severe tinnitus and vertigo. Two patients described auditory hallucinations in the affected ear and

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these were described as voices. No deafness was noted in four patients but two of these had an absent caloric response.

Vestibular function was assessed in 180 patients by the response to an iced water caloric test. Only one patient showed a normal vigorous response to this maximal stimulus.

TABLE II.

AUDITORY FINDINGS IN 214 PATIENTS

Deafness and tinnitus	124
Deafness alone	82
Tinnitus alone	2
Unco-operative	2
No deafness	4

Headache

Headache was present in 156 patients and occurred mainly in the occipital or frontal regions (Table III). The relationship between headache, papilloedema and pressure upon the Trigeminal nerve has been discussed by Edwards and Paterson (1951) and no positive conclusions were drawn. We found that there was no constant relationship between the presence or site of headache and the presence of papilloedema or Trigeminal nerve involvement in this series.

TABLE III.

SITES OF HEADACHE IN 214 PATIENTS

Occipital	49
Frontal	42
Fronto-occipital	24
Occipito-frontal	8
Vertex	18
Facial pain	10
Otalgia	5
None	58
Total with headache	156 (73 per cent)

Cerebellum

Nystagmus and limb inco-ordination were taken as signs of cerebellar involvement. In 168 cases the nystagmus was coarse when looking to the side of the lesion and fine when looking away from it, while in twenty-five cases this was reversed. In twenty-one patients there was no observable nystagmus. One-hundred-and-seventeen patients showed limb inco-ordination which was ipsilateral in 111 cases, contralateral in four cases; and two pstienta had involvement of all four limbs.

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Cranial neuropathy

There were no oculomotor or trochlear nerve palsies noted in this series. Twenty-four patients had an ipsilateral abducent nerve palsy and a further twelve had bilateral palsies.

There was trigeminal nerve involvement in 179 patients. Of these, 115 showed diminution in sensitivity of the corneal reflex only, sixty-two had hemifacial anaesthesia and twenty-four cases showed masseteric weakness (Table IV). Six patients had bilateral Vth-nerve signs and in one patient there was contralateral nerve involvement alone.

TABLE IV.

CRANIAL NERVE SIGNS IN 214 PATIENTS

Cranial nerve	Sign with numbers	Percentage
V	Reduced corneal reflex only (115) Hemifacial anaesthesia (62) Masseteric weakness (24)	83
VI	Ipsilateral (24) Bilateral (12)	17
VII	Complete facial palsy (29) Partial facial palsy (107)	64
IX, X, XI, XII	Signs present (22)	10
Long tract signs	Signs present (16)	7

Twenty-nine patients had a total facial palsy, 107 patients had incomplete palsies and seventy-eight had no evidence of facial weakness in the immediate preoperative period. Twenty patients had hemifacial spasm.

Palsies of the IXth, Xth, XIth and XIIth cranial nerves were uncommon in the series and were seen in twenty-two patients. These nerves tended to be involved together and to be accompanied by dysarthria, dysphagia and aspiration pneumonia.

Raised intracranial pressure and medullary compression

Papilloedema was present in 150 patients of whom sixty-eight had failing vision and six were blind. Seventeen patients were anosmic. Ten patients had epileptiform convulsions and in two of these there were features typical of cerebellar fits, namely associated respiratory and circulatory disturbances. Ventricular pressure was measured at craniotomy in ninety-six cases and in fifty-eight of these the pressure was above 170 mm. of cerebrospinal fluid. Long tract signs such as spasticity or sensory deficit were seen in sixteen patients and indicated a poor prognosis.

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Treatment

A total of 202 patients were operated upon by the posterior fossa sub-occipital craniotomy route. Partial removal of tumour was achieved in 121 patients and thirty-six of these required further surgery for residual tumour. Seventy-three patients had total removal of tumour and eight patients had a decompression operation only, because of their poor general condition.

Mortality and morbidity

There were thirty-four operative deaths in the series, a mortality of 17 per cent overall. This compares favourably with other figures for large tumours (Revilla, 1947). The commonest cause of death was brain stem injury or compression due to injury to vital arteries, attempts to free adherent tumour or postoperative bleeding. Three patients developed meningitis early in the postoperative period and two patients died with aspiration pneumonia due to bulbar palsy.

One-hundred-and-fifty-two patients were considered to have a severe physical handicap in the early postoperative period. One-hundred-and-seven had increased cerebellar ataxia with marked limb inco-ordination, but many were lost to long-term follow-up and the number with permanent ataxia is not known. However, of fifty-seven male long-term survivors, only fifteen returned to non-sheltered gainful employment.

Eleven patients had total loss of sight in both eyes and eighteen patients had severely-impaired visual acuity, due to secondary optic atrophy or neuroparalytic keratitis. Ten patients had swallowing inco-ordination with aspiration of ingested food and consequent recurrent chest infections while six patients had a profound contralateral hemiplegia.

Total facial palsy was present in all those cases of total tumour removal as curettage and diathermy to the internal auditory meatus was performed. In those cases where partial removal was performed twelve patients retained facial nerve function.

Discussion and summary

It was formerly held that surgery for acoustic neuroma should be delayed until signs of raised intracranial pressure or severe physical disability were apparent (Pennybacker and Cairns, 1950), as it was thought that many patients with minimal symptoms may continue in good health for many years. However, in this series the average period between the onset of symptoms and significant neurological deficit necessitating surgical relief was four-and-a-half years. This was a surprisingly short time interval.

Although the overall operative mortality for these large tumours was low, there was a considerable degree of morbidity. Long-standing and progressive raised intracranial pressure led to secondary optic atrophy and

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blindness. An additional hazard to sight in these cases was a combination of corneal anaesthesia and inability to close the eye leading to neuroparalytic keratitis. Lateral tarsorrhaphy and fascial sling operations were only partially successful in preventing this complication.

Loss of motor function whether due to cerebellar ataxia or hemiplegia was a severe handicap and was especially serious in those young patients who had continuing family responsibilities. These patients required continued supportive care from hospital departments, family doctor and local authority services.

Techniques for the total removal of small and medium-sized tumours via the translabyrinthine and middle fossa routes have now been developed (House, 1968; Morrison and King, 1973). These approaches are associated with minimal postoperative morbidity. The inevitable mortality and morbidity associated with the surgical removal of large tumours adherent to the brain stem can only be avoided by earlier diagnosis and earlier treatment.

Sophisticated audiometric and neuroradiological techniques are now available in most large centres and these enable an early diagnosis to be made if a high index of clinical suspicion is maintained by practising otolaryngologists.

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Reprints

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