The bloody angle: 100 years of acoustic neuroma surgery

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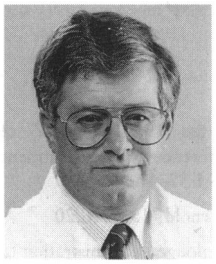
J R Soc Med 1995;**88**:464P–468P

PRESIDENTIAL ADDRESS READ TO SECTION OF OTOLOGY, 4 NOVEMBER 1994



Professor Richard Ramsden has recently been appointed Honorary Visiting Professor in Otolaryngology at the Victoria University of Manchester. He has been Consultant Otolaryngologist at Manchester Royal Infirmary since 1977. His special interests are in all aspects of otology, but in particular the fields of neuro-otology and skull base surgery in which he has built up a multidisciplinary team approach to the management of cerebellopontine angle tumours. He was one of the first in the UK to realize the potential of cochlea implantation in the rehabilitation of the profoundly deaf, and leads a large adult and paediatric implant programme. He was President of the Section of Otology 1994–1995.

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Keywords: *acoustic neuroma; vestibular schwannoma; history of medicine; tumours*

As far as one can tell the earliest evidence for the presence of eighth nerve tumours comes from the archaeological excavations in a graveyard in Franzhausen in Austria dating back to 2500 BC. Pirsig *et al.*[1] describe the findings from two children whose temporal bones were examined and then scanned. In both, there was marked widening of one internal auditory meatus and in one there were changes to suggest extension of tumour into the cochlea. These authors suggest that these two patients suffered from neurofibromatosis type 2 (NF2).

The first fully documented postmortem description of an acoustic neuroma dates back to 1777 to Eduard Sandifort[2] who was professor of pathology in Leiden. His description 'De duro quodam corpusculo, nervo auditorio adherente' is a classic. The 'certain hard body' was

> not only connected to the lower part of the said nerve, but also adhered to the nearest part of the medulla oblongata from which the two seventh nerves emerge, penetrating also as far as the foramen in the inner part of the petrous section of the temporal bone.

He described the appearance of the small growth, externally hard like cartilage but internally quite soft. He pointed out that this cause of deafness was clearly beyond the reach of medication or surgery and must be declared incurable. His conclusion was that tumours such as he had described could produce deafness, in the manner of 'effusions of the humours, disease and bony exostoses'.

During the early nineteenth century a number of case reports appeared in which antemortem symptoms and clinical signs were correlated with postmortem findings. The earliest was probably that of Leveque Lasource in the French literature in 1810[3]. The earliest to appear in the English literature was Sir Charles Bell's[4] case of a young woman who presented to him in 1830 with anaesthesia of the lower two divisions of the left trigeminal nerve so profound that:

> the end of a feather passed three inches into the nostril gives her no sensation, and does not produce sneezing.

She subsequently developed a leftsided hearing loss and facial paralysis, giddiness, headache and vomiting. Bell describes the final stages of her brain stem failure with clenching of the teeth, indistinct speech and failing respiration and swallowing. At post mortem a tumour 'about the size and not unlike the form of a pigeon's egg was discovered on dividing the tentorium'. The tumour was cystic and contained fluid the colour of urine. The solid portion had a consistency similar to vitreous humour. The tumour filled the cerebellopontine angle, indented the pons and the cerebellum and extended into the internal meatus. No recognizable nerve could be seen entering the internal meatus, but the medial $^1/_4$ in. of the facial nerve could be seen leaving the stem. Only the medial $^1/_2$ in. of the trigeminal nerve could be identified before it too was lost in the tumour.

There are many more fascinating accounts from these masterly nineteenth century observers, in particular that of Cruveilhier[5], in 1835, whose report cannot be bettered as an account of the progression of the symptomatology of acoustic neuromas from deafness to postmortem. It is clear that at that time diagnostic skills were progressing at a greater rate than therapeutic remedies. Cruveilhier describes the application of 16 leeches to the mastoid, blood letting from the feet, electropuncture and the use of moxas. A moxa is downy vegetable material used in Oriental medicine as a counterirritant by igniting it on the skin.

Debate also began regarding the site and cell of origin of these growths. The term acoustic tumours soon became fairly commonplace, but histologically they were variously referred to as fibrosarcomata, gliofibromata, neuromata and neurofibromata. Rokitanski remarked in 1848 that neuromas could occur on all cranial nerves except the olfactory, optic and acoustic nerves. It was not until much later that the true origin from the Schwann cells was to become apparent, although Theodore Schwann who was professor of anatomy and physiology in Liège described the nerve sheath in 1838.

Accurate preoperative diagnosis gradually developed as clinical syndromes were correlated with postmortem findings, but it was not until the last 20 years of the nineteenth century that localization of a tumour could be predicted with sufficient confidence to advise surgery. This increased accuracy owed much to the work of Hughlings Jackson and Gowers in England, Bruns and Oppenheim in Germany, and Babinski in France in defining the features of posterior fossa and particularly cerebellar pathology.

There must have been a number of unreported unsuccessful attempts at tumour removal during the latter half of the nineteenth century. A case was operated on by von Bergmann in Oppenheim's clinic in 1890, but the tumour was not found until after the patient's death, an event which ensued fairly quickly. Ballance in London is widely, but probably erroneously, credited with the first successful removal of an acoustic neuroma in 1892. The case note records that the tumour was widely attached to the dura of the posterior surface of the petrous bone, and, furthermore, there was no mention of deafness as a symptom. It seems more likely, as stated by Cushing[6], that this tumour was, in fact, a meningioma. Cushing himself attributed the honour to Thomas Annandale, Professor of Surgery in Edinburgh whom he credits with 'a brilliant surgical result, the first recorded' almost exactly a century ago on 3 May 1895.

Thomas Annandale was born in Newcastle upon Tyne in 1838 and studied medicine in Edinburgh where he eventually succeeded Lister as Regius Professor in 1877. Like all general surgeons of the day, he truly could turn his hand to anything, although most of his practice seems to have been orthopaedic. His MD thesis for which he was awarded Highest Honours and the Gold Medal was 'On the injuries and diseases of the hip joint'. His famous case described by Gibson (1896)[7] was a pregnant young lady from Dundee with a rightsided hearing loss, evidence of lower cranial nerve involvement, a central pattern of nystagmus, long tract signs, contralateral false lateralizing signs and papilloedema, i.e. strong clinical evidence of a large posterior fossa tumour. After ineffective treatment with inunction of blue ointment, Annandale trephined the skull over the right lobe of the cerebellum and removed a semicystic tumour the size of a pigeon's egg. Microscopic investigation showed it to be of the nature of a fibrosarcoma, at the time a rather imprecise term, but from the clinical description of the case it must have been an acoustic neuroma. Postoperatively progress was satisfactory and the patient returned home and subsequently gave birth to a healthy child. She appears to have suffered no significant neurological deficit from the surgery.

Despite Annandale's success, surgical results at the turn of the century were ominously dreadful. The name of Krause has become associated with the unilateral suboccipital approach. Various proponents of the technique reported their results for attempted acoustic neuroma removal, and the procedure which entailed the digital enucleation of the tumour with emphasis on celerity of execution, was accompanied by 'a shocking mortality' in the words of Cushing. Borchardt reported an operative mortality of 72%, von Eiselberg 74% and Krause himself 84%[8]. One of the reasons for the dire results was the failure of surgeons to appreciate that the anterior inferior cerebellar artery was essential for the nutrition of the brain stem and that damage to it, by the enucleating finger, was usually fatal. Ballance indeed suggested that tumour removal would be so much easier if only the artery could be ligated first. Remarkably, the translabyrinthine approach was first proposed by Panse[9] in 1904 as a response to the terrible morbidity associated with the Krause operation. Panse himself felt that the access would be restricted and that only the smallest tumours would be removable through this approach. Furthermore, loss of the facial nerve was regarded as inevitable and complete tumour removal was considered unlikely. Operations were carried out by Kummel in Heidelberg in 1909 and by Quix[10] in Utrecht in 1911 but with limited success because of cramped access. Ballance was pretty dismissive of the approach when he described it as 'objectional for obvious reasons'.

The bilateral operation first introduced by Harvey Cushing in 1905 was advocated for the exploration of obvious subtentorial lesions not definitely localized to one side or the other: the idea being that if the tumour was not found in the first cerebellopontine angle, the other side could easily be examined through the same approach. It was Cushing who, recognizing the dire problems of haemorrhage during acoustic neuroma surgery, compared the cerebellopontine angle with the fence corner at the Battle of Gettysburg and suggested that it might well be called the 'bloody angle'.

Cushing was without doubt one of the most remarkable men to have graced surgery. He established the identity of the neurosurgical specialist and drew up the rules of neurosurgical practice and procedure which persist to this day. His meticulous attention to detail as a surgeon, and as a recorder and reporter of his work established a phenotype from which several generations of neurosurgeons have been

cloned. His practice embraced all aspects of neurosurgery but he will be particularly remembered for his contributions to surgery of the pituitary gland which led, of course, to his major endocrinological work and the description of the disease that bears his name. His other great contribution was in the surgery of acoustic neuromas. His monograph *Tumors of the Nervus Acusticus and the Syndrome of the Cerebellopontile Angle*, published first in 1917 is a classic and should be compulsory reading for all those with an interest in this subject. In this work, he was able to demonstrate how, by employing the technique of intracapsular debulking, he had been able to reduce the perioperative mortality first to 35% and then to 10%. In addition to having great surgical skills Cushing was also a formidable writer, and bibliophile. His collection of old medical texts was unique, particularly the works of Vesalius. Cushing was awarded the Pulitzer Prize for his biography of Sir William Osler, and curiously Fulton also won that prize for his biography of Cushing. Cushing's results were a dramatic improvement on those of all those who had gone before. Nevertheless, his subtotal removal inevitably resulted in a high number of recurrences, and his great rival of the time Walter E Dandy[11] soon espoused the philosophy of total tumour removal through a unilateral suboccipital approach. His results too were good but despite the advances of these two great Americans, neurosurgeons by and large remained somewhat daunted by the prospect of operating on these tumours and were reluctant to recommend surgery until the tumours were very large. This approach, of course, had the effect of perpetuating itself. Do not operate on tumours until they are large, because the results are bad. Operate on them when they become large and the results are indeed bad. QED. Attitudes to surgery as late as the 1950s are encapsulated in the oft quoted report of Pennybacker and Cairns[12]. They reported a perioperative mortality of 20% in a series of 130 cases almost all of which had pretty massive tumours at the time of surgery, which in most instances comprised intracapsular debulking. Northfield[13], writing at the same time, reported a perioperative mortality of 38%. In the series of partial removals reported by Givre and Olivecrona in 1949[14] 60% of patients were dead of recurrence in 4 years. In Northfield's 1970 series reported at the Royal Society of Medicine the average tumour size was 3–4 cm, and the perioperative death rate was still 16%, but he now recommended total, early removal of the tumour at the first attempt, albeit with disregard for the facial nerve which he saw as being of secondary importance[15].

The stage was set for the arrival of William House, who in 1961 presented his candidate's thesis to the American Laryngological Rhinological and Otological Society in which he described the microsurgical exposure of the internal auditory canal and its contents through the middle cranial fossa, and suggested it might be suitable for removal of acoustic tumours[16]. It is interesting that one of House's reasons for developing the approach was to remove otosclerotic foci from the internal meatus where they were thought to cause pressure effects on the eighth nerve. Two years later House reported 10 cases of acoustic tumours removed by the middle fossa approach and 10 removed by the translabyrinthine approach[17].

The response from the neurosurgeons in the audience, Norman Dott and Charles Drake, was remarkably positive. There was little to suggest the vitriolic exchanges between the neurosurgeons and the otologists that were to follow. The neurosurgeons were particularly critical of what they saw as being very limited access through the labyrinth and the high risk of cerebrospinal fluid leakage. House and his followers pointed out the fact that cerebellar retraction, or indeed resection, were unnecessary, and that with certain modifications any size of tumour could be removed through the petrous bone. In addition, early identification of the facial nerve allowed preservation to be the rule rather than the exception. Particular mention must be made of Bill Hitselberger, who was branded a pariah in neurosurgical circles for daring to agree and work with House. Eventually, however, the message began to emerge that results of acoustic tumour surgery were likely to improve if the combined talents, techniques and philosophies of the two surgical specialities were harnessed for the common good of the patient, and combined neurotological teams are now the rule where this work is performed.

Advances in diagnosis were of great importance too. In the period just before and after the Second World War tests of audiological function were introduced which it was hoped would allow surgeons to differentiate between neural and sensory (i.e. cochlear) deafness. Fowler's alternate loudness balance test, Carhart's test for tone decay, Bekesy audiometry, the loudness discomfort test, the short increment sensitivity index (SISI) and speech audiometry were in the first generation and later came tests of aspects of the stapedial reflex, its threshold and its decay characteristics. These tests were seen as a great breakthrough and allowed otologists to speculate with variable degrees of accuracy as to whether a tumour was present. Their reliability was, however, not good and undue dependence upon them probably led to a large number of missed diagnoses. The audiological test which we are led to believe is the most accurate is the auditory brainstem response (ABR), with a quoted sensitivity of 98%. This certainly sounds impressive until one realizes that the specificity of the test is very low indeed. Any hearing loss greater than 70 dB whether cochlear or retrocochlear, will be associated with an ABR abnormality, i.e. absence of response, which is due to the severity of the hearing loss itself rather than the site of the lesion. It is certainly true that abnormalities of the ABR will be found in almost all proven

cases of acoustic neuromas, although if so much credence is placed upon the ABR that further investigation ceases on obtaining a normal response, this will lead to the anomaly of the self fulfilling prediction, that is to say the cases with the negative ABR will *ipso facto* be deemed to be tumourless. It is in the field of imaging that advances have been most spectacular. In the space of 20 years we have progressed from the era of the Technesium 99 scan which could only be relied upon to detect lesions of approximately 3 cm or larger, to the latest generation of magnetic resonance imaging (MRI) which with gadolinium enhancement can demonstrate intrameatal lesions of 2 mm or less. Plain radiology, petrous tomography, myodil meatography and air meatography have all come and all have gone. Even computerized tomography scanning, without intrathecal contrast, cannot be relied upon to detect a tumour with an intracranial diameter of less than 1.5 cm.

The current prospects for most patients undergoing acoustic tumour surgery are: total tumour removal; perioperative mortality risk of under 1%; a risk of major neurological sequelae of approximately the same magnitude; a 95% chance of facial nerve preservation; 7–10 days in hospital; and 2 months off work and return to a normal level of physical activity. The two factors which have been shown over and over again to be the most important in predicting outcome are tumour size and experience of the operating team. Facial nerve preservation does not mean normal facial function, of course, but again the smaller the tumour and the more experienced the surgeon the higher the percentage of Grade 1 and 2 results.

The picture at the end of the first 100 years of acoustic neuroma surgery is thus a much brighter one than when Annandale achieved his notable first success. There are, nevertheless, always new issues to confront the otoneurosurgeon dealing with these fascinating tumours. Listed below are some of what are certainly some of the most contentious, many of which are interrelated.

Current issues

(i) What do we do with the early diagnosed cases from MRI?
(ii) Can tumour growth characteristics be predicted?
(iii) What is the value of hearing preservation surgery?
(iv) Does stereotactic radiosurgery have a place in treatment?
(v) What advances are there in the management of NF2?

MRI now allows us to detect the smallest of tumours. There is no certainty that all of these early tumours need to be removed, and factors such as hearing preservation and tumour growth rate need to be considered. Despite the current enthusiasm for hearing preservation surgery there is no doubt that at the present time the best chance of preserving hearing is not to remove the tumour. This policy is acceptable if the tumour is small, the hearing is of value to the patient and the patient agrees with the management policy which requires a commitment to attend regularly for followup scanning. Acoustic tumours do not all grow at the same rate, and it would be valuable to be able to identify some sort of marker in the bloodstream or perhaps in biopsy material that would help predict future growth. One could, perhaps, envisage a situation in which a tumour could be biopsied through a bur hole and its growth characteristics established by a simple biological test. A number of studies have looked at the S phase percentage, at Ki67 monoclonal antibodies, at ribosomal RNA concentrations, and at proliferating cell nuclear antigen without any convincing association with tumour growth. Any treatment policy based on such a strategy would presuppose that growth rate remained linear but there is some evidence that this is not necessarily the case.

Controversy surrounds the use of stereotactic radiosurgery (the gamma knife) and space does not allow a full examination of the issues. Suffice it to say that most surgeons remain unconvinced by its proponents, but recognize the need to evaluate claims in a dispassionate manner as results are published.

The exact definition and nature of NF2 has been clarified by recent advances in molecular biology by Troffater *et al.*[18] with identification of the tumour suppressor gene for NF2 and indeed for sporadic unilateral acoustic neuromas on the q arm of chromosome 22. Furthermore, MRI of the spine as well as the brain has allowed more accurate staging of NF2 and enabled surgeons to plan treatment with more precision. Hearing preservation surgery has a definite place in the management of NF2, and when it is unsuccessful, advances in microelectronics and preoperative monitoring have allowed auditory rehabilitation of some individuals using either cochlear implants or the auditory brain stem implant. One imagines that it will only be matter of time before correction of the molecular abnormality becomes a practical proposition. It seems that the first century of acoustic neuroma surgery could well be the last. In a sense 'acoustic neuroma' surgery is already a thing of the past. As the second century dawns, in accordance with the edicts of the National Institutes of Health (Bethesda, MD, USA), it has now, correctly, to be replaced by the more pathologically accurate 'vestibular schwannoma' surgery.

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(Accepted 21 March 1995)