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Clinical correlates of acoustic neuroma morphology

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Abstract
Thirty-eight patients with vestibular schwannomas were reviewed. A correlation was found between tumour morphology and clinical presentation. Based on our knowledge of the variability in the neurilemmal–neuroglial junction and therefore the site of origin of these tumours in relation to the internal auditory canal, a classification into three different appearances on magnetic resonance imaging was possible. Dumbbell shaped tumours (21 per cent) represented laterally arising schwannomas, lollipop shaped tumours (18 per cent) were medially arising and cone shaped tumours (61 per cent) were the more common intermediate form. Patients with laterally arising dumb-bell shaped tumours were more likely to present early with hearing loss and had smaller tumours than patients with medially arising lollipop shaped ones. The relatively well preserved hearing in patients with medially arising tumours made them more likely to present at a later stage with signs of trigeminal compression, cerebellar dysfunction and raised intracranial pressure.

Key words: Cranial nerve neoplasms, vestibular schwannoma; Classification

Introduction

The vestibular nerve is surrounded by a proximal neuroglial and a distal neurilemmal or Schwann cell sheath (Nager and Baltimore, 1969). Vestibular schwannomas originate in the distal neurilemmal portion of the vestibular nerve at or close to the neurilemmal–neuroglial junction (Skinner, 1929). They are benign neoplasms of the Schwann cell sheath. The position of the neurilemmal–glial junction and thus the site of origin of these tumours will vary. Some vestibular schwannomas are medially arising with little or no intracanalicular component while others are laterally arising with a large intracanalicular component (Luetje *et al.*, 1983). The proportion of tumour within the internal auditory canal (IAC) varies and less commonly the schwannoma arises medial to the porus acousticus in the cerebello-pontine angle (CPA) and has no intracanalicular component. It might be expected, therefore, that the degree of compression of the contents of the IAC and thus the clinical and audiological presentation of medially and laterally arising tumours may vary (Moffat *et al.*, 1989a; Baguley *et al.*, 1989).

Magnetic resonance imaging (MRI) is now the radiological method of choice for investigating suspected vestibular schwannomas (House *et al.*, 1986; Mikhael *et al.*, 1987). It gives better soft tissue contrast than computed tomography (CT) (Bradley and Shelden, 1983) and is particularly useful in visualizing the contents of the IAC since bone has a negative signal (Kingsley *et al.*, 1985; Daniels *et al.*, 1987).

The aims of this study were to differentiate the morphological types of acoustic neuroma on MRI scanning and to correlate this with the differences in clinical and audiological presentation in medially and laterally arising tumours.

Materials and methods

The medical records and MRI scans of 38 patients who had histologically proven vestibular schwannomas excised by the senior authors (D.A.M., D.G.H.) in the Department of Otoneurosurgery, Addenbrooke's Hospital, Cambridge between January 1990 and December 1991 were reviewed.

A detailed history was taken from each patient and the first symptom was noted. A complete neuro-otological examination was carried out. Every patient had a pure tone audiogram (PTA) as a part of the audiovestibular investigation which also included a speech audiogram, a caloric

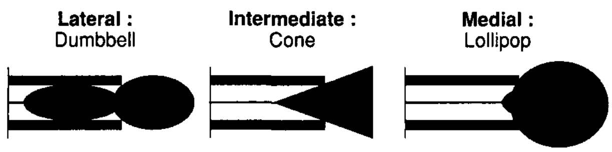

FIG. 1

Diagram to illustrate the form of dumbbell, cone and lollipop shaped vestibular schwannomas. Dumbbell tumours arise laterally; cone shaped tumours arise near the porus; lollipop shaped tumours arise medial to the porus.

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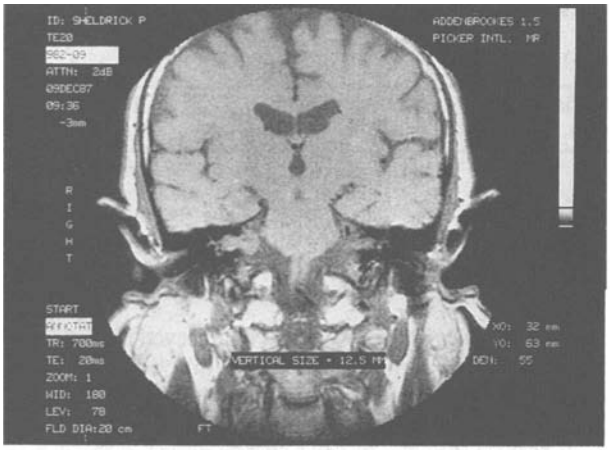

FIG. 2

Magnetic resonance imaging scan of a dumbbell shaped vestibular schwannoma: in coronal view. The neurolemmal–neuroglial junction is lateral to the porus acousticus.

test and auditory brainstem responses (ABR). The mean hearing loss was calculated as the average of 500, 1000, 2000 and 4000 Hz thresholds. All patients had a preoperative MRI scan. This excluded a number of patients operated on between January 1990 and December 1991 who had only undergone a pre-operative CT scan.

The MRI scans were reviewed by consultants in neuro-otology and neuro-radiology. Each patient had both axial

FIG. 3

Magnetic resonance imaging scan of a cone shaped vestibular schwannoma: in coronal view. The neurolemmal–neuroglial junction is near the porus acousticus.

FIG. 4

Magnetic resonance imaging scan of a lollipop shaped vestibular schwannoma: in coronal view. The neurolemmal–neuroglial junction is medial to the porus acousticus.

and coronal views. It was possible to classify each tumour into one of three groups: dumbbell shaped, cone shaped and lollipop shaped (Fig. 1).

Dumbbell shaped tumours filled the IAC, expanding it near its mid-point. The 'waist' of the dumbbell was at the porus acousticus with the tumour expanding out into the CPA (Fig. 2). Cone shaped tumours had a more medially placed intracanalicular component which 'funnelled' the porus before expanding out into the CPA (Fig. 3). Lollipop shaped tumours had a very small or no intracanalicular component which along with the VIIth and VIIIth nerve complex made up the 'stick' of the lollipop (Fig. 4), most of the tumour being confined as a spherical or ovoid mass in the CPA.

The maximum intracranial diameter of each tumour was computed on the MRI scanner programme. The schwannomas were divided into four groups by size: small tumours measured less than 10 mm in greatest diameter; medium sized tumours between 10 mm and 20 mm; large tumours 20 mm to 30 mm; and giant tumours were greater than 30 mm.

Results

Tumour morphology

Thirty-eight MRI scans were analysed and it was possible to classify each schwannoma into one of three groups. Twenty-three (61 per cent) of the tumours were cone shaped, eight (21 per cent) were dumbbell shaped

TABLE I

DEMOGRAPHICS OF PATIENTS WITH DUMBBELL, CONE AND LOLLIPOP SHAPED VESTIBULAR SCHWANNOMAS

	Cones (N = 23)	Dumb-bells (N = 8)	Lollipops (N = 7)	Total (N = 38)
Mean age	51	47	47	50
Female:male	13:10	4:4	4:3	21:17
Side right: left	11:12	3:5	2:5	16:22
Mean length of history (years)	3	2	6	4

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TABLE II
TUMOUR SIZE (MAXIMUM DIAMETER)

Size of tumour (Diameter)	Cones (N = 23)	Dumbbells (N = 8)	Lollipops (N = 7)	Total (N = 38)
Small (<10 mm)	3	2	0	5
Medium (10–20 mm)	13	5	3	21
Large (20–30 mm)	7	1	1	9
Giant (30–40 mm)	0	0	3	3
Mean diameter in mm	15	12	24	16

and seven (18 per cent) were lollipop shaped. The mean age and sex distribution of the three groups were comparable, although the mean length of history was longer for the patients with lollipop shaped tumours (Table I).

Tumour size

There were five (13 per cent) small tumours, 21 (55 per cent) medium sized tumours, nine (24 per cent) large tumours and three (8 per cent) giant tumours. A significant association was discovered between tumour size and shape. Nearly 90 per cent of the dumbbell shaped schwannomas were small or medium sized, compared with 70 per cent of the cone shaped tumours. In the case of the lollipop shaped tumours 60 per cent were large or giant sized (Table II). A comparison of the mean greatest diameter using the chi-square test (Petrie, 1978) revealed that the lollipop group were significantly larger than both the dumbbell ($p<0.001$) and the cone shaped tumours ($p<0.01$).

Symptoms

The initial presenting symptoms in the 38 patients in this series can be seen in Fig. 5. All eight patients with dumbbell shaped tumours noted hearing loss as their first symptom. In contrast, 15 (65 per cent) of patients with cone shaped tumours and only three (40 per cent) of patients with lollipop shaped tumours presented with hearing loss. The first symptom in 30 per cent of patients with lollipop shaped tumours was decreased facial sensation.

Signs

A depressed corneal reflex, the earliest sign of trigeminal nerve compression, was seen in all seven patients with lollipop shaped tumours. However, this was the case in only two (25 per cent) patients with dumbbell shaped tumours. Similarly, decreased facial sensation was evident in four patients (60 per cent) with lollipop shaped schwannomas but in only one (5 per cent) patient with a cone shaped tumour and in none of the patients with dumbbell shaped tumours. As might be expected from the large size and medial position of lollipop shaped tumours, tests of cerebellar dysfunction demonstrated abnormal gait and upper limb ataxia and was more frequently seen in these patients than in the other two types (Table III).

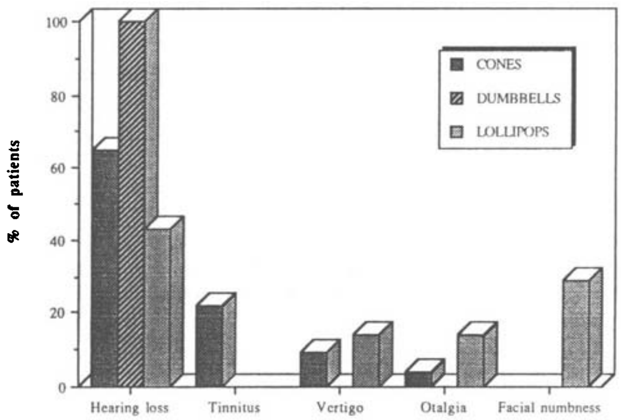

FIG. 5

Histogram illustrating the initial presenting symptom of 38 patients with different types of vestibular schwannoma.

Pure tone audiometry

Despite the large size of their schwannomas, patients with lollipop shaped tumours had much better preserved hearing than the other two groups (Fig. 6). All seven cases had a hearing loss of less than 60 dB and one patient had normal hearing. In contrast, six (75 per cent) of patients with dumbbell shaped tumours had a hearing loss of greater than 80 dB. Statistical comparison of the mean hearing loss (= average hearing threshold at 500, 1,000, 2,000, 4,000 Hz) using chi-square test revealed that: (1) the mean hearing loss of each group was significantly different; (2) patients with dumbbell shaped tumours had a much more profound hearing loss than lollipop shaped schwannomas ($p<0.001$); (3) hearing deficit in patients with cone shaped tumours was intermediate: it was better than the dumbbell group ($p<0.01$) but significantly worse than that of the lollipop group ($p<0.001$).

Discussion

As a vestibular schwannoma grows it will produce sequential symptoms and signs. The evolution of these have been divided into four stages (Ramsden, 1987). Firstly, the otological stage which is usually characterized by ipsilateral tinnitus, high frequency sensorineural hearing loss and dysequilibrium. This represents involvement of the vestibular and cochlear nerves with gradual impairment of function. Sudden sensorineural hearing loss occurs in 10 per cent of patients perhaps by tumour compression of the internal labyrinthine artery in the IAC (Moffat *et al.*, 1989a).

As the tumour grows out from the porus acousticus it will begin to touch the trigeminal nerve when it is 2 cm in size and this is the second stage. In the third stage the tumour will fill the CPA and begin to compress the brainstem and cerebellum producing cerebellar dysfunction. Finally, shift of the fourth ventricle and obstruction of the cerebrospinal fluid pathways will result in hydrocephalus, raised intracranial pressure and papilloedema. Decompensation with respiratory depression due to irretrievable

TABLE III
NEURO-OTOLOGICAL SIGNS

Sign	Cone (N = 23)	Dumb-bell (N = 8)	Lollipop (N = 7)	Total (N = 38)
Sensory-neural hearing loss	21	8	5	36
Decreased corneal reflex	3	2	7	12
Decreased facial sensation	1	0	4	5
Nystagmus	2	0	2	4
Papilloedema	0	0	1	1
Gait:*				
W.A.L.E.O.	7	1	5	13
W.A.L.E.C.	18	5	7	30
Unterberger positive	11	6	5	22
Romberg positive	0	2	2	4
Upper limb ataxia	0	0	2	2

*W.A.L.E.O. = walking along a line with eyes open (principally a test of cerebellar function). W.A.L.E.C. = walking along a line with eyes closed (principally a test of vestibular function).

brainstem compression of the respiratory and cardiovascular centres will occur as a terminal event.

Vestibular schwannomas occur at the neurilemmal–glial junction and although this is usually in the region of the porus acousticus, its site varies considerably. Thus tumours may arise laterally and fill the fundus of the IAC, they may arise at the porus acousticus or medially in the CPA. The laterally arising tumours tend to be dumb-bell in shape, the porus acousticus producing the waist of the dumb-bell. Intermediate tumours may be cone shaped, the porus acousticus and medial part of the canal being expanded as the tumours grows out into the CPA. Medially arising tumours originate within the CPA and the IAC is not expanded and may not contain any tumour.

In this study there was a clear correlation between tumour morphology and hearing loss. The patients with laterally arising dumbbell shaped tumours had a severe hearing deficit. In comparison, patients with medially arising lollipop shaped tumours had significantly better preserved hearing ($p<0.001$). The hearing in medially arising tumours was also significantly better preserved than patients with intermediate or cone shaped tumours ($p<0.001$).

The audiogram results must also be related to tumour size, since this is known to have an important correlation

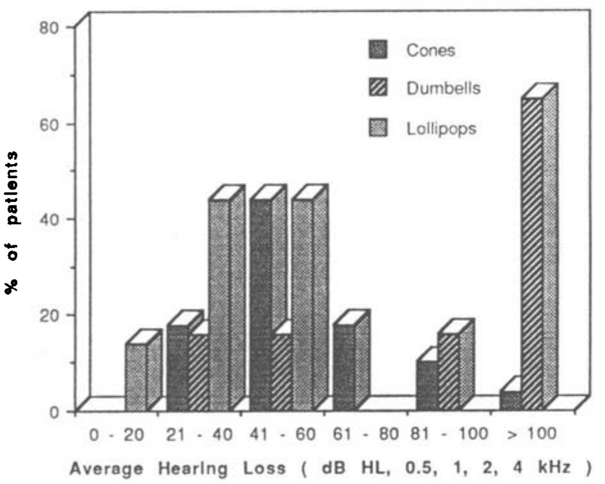

FIG. 6

Histogram illustrating the average hearing loss for 38 patients with different types of vestibular schwannoma. (Hearing loss is averaged over four frequencies: 0.5, 1, 2 and 4 kHz; and patients are grouped into 20 dB bands.)

with hearing loss (Thomsen and Tos, 1990). The analysis of tumour size in this study, as in most studies to date (Kasantikul *et al.*, 1980), was based on maximum tumour diameter. Medially arising tumours were significantly larger than laterally arising ($p<0.001$) and intermediate ($p<0.01$) tumours which makes the fact that the hearing is better preserved in these tumours even more significant.

It seems probable that the first otological stage of tumour growth is produced by compression of the neural structures within the bone of the IAC. This leads to dysequilibrium and loss of vestibular function, tinnitus and gradual sensorineural hearing loss. The poor speech discrimination seen in these patients may result from this compression with its concomitant effect on the blood supply of the cochleovestibular apparatus and VIIth and VIIIth cranial nerves. Sudden occlusion of the internal labyrinthine artery may be responsible for the sudden deafness seen in 10 per cent of these patients (Moffat *et al.*, 1989a). Medially arising tumours have the space to grow in the CPA without this compressive effect and this could account for the less marked or even absent otological stage and the tendency to present later with symptoms and signs related to trigeminal and cerebellar dysfunction.

In a previous study (Baguley *et al.*, 1989) we reported transtympanic electrocochleographic (ECochG) findings in medially and laterally arising acoustic neuromas. The AP/SP complex was more likely to be normal in medially arising tumours whereas the abnormal widened waveform (Kiang Potential) reported by Morrison *et al.* (1976) was more likely to be seen in laterally arising tumours due to the compressive effect of the tumour on the cochlear nerve in the IAC. Also a significantly higher proportion of patients with medially placed tumours had thresholds by ECochG better than by PTA when compared with laterally arising tumours. Daumann *et al.* (1988) noted that the N1 by ECochG arises from the cochlea and lateral part of the VIIIth nerve which could explain these findings.

The early detection of acoustic neuroma is of paramount importance if the morbidity of the surgery of these tumours is to be reduced further (Moffat *et al.*, 1989a,b). MRI scanning with gadolinum DTPA enhancement will demonstrate tumours as small as 2 mm. Despite this and the advances in audiovestibular investigative techniques some tumours continue to present at a late stage. By correlating the presenting symptoms and signs with the tumour morphology this paper explains why some tumours, particularly those which are medially arising, are more likely to present late.

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