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Main Article

Diagnostic delays in vestibular schwannoma

H Teppo, J Heikkinen*, K Laitakari*, O-P Alho*

Abstract

Objectives: The literature on delays in vestibular schwannoma diagnosis is from the era before the routine use of magnetic resonance imaging. We evaluated such diagnostic delays and their impact on tumour size and on pre- and post-treatment morbidity, in a relatively recent patient series.

Study design: Retrospective review.

Methods: A two-centre study was conducted, including 91 consecutive vestibular schwannoma patients diagnosed between 1992 and 2006. Data on the presenting symptom and the initial medical visit were obtained from primary care records completed at the time of the initial visit; data on the tumour and the clinical course were obtained from review of the hospital chart. Data on diagnostic delays were available for 59 patients.

Results: The median patient, professional and total diagnostic delays were three, four and 14 months, respectively. Unilateral hearing loss as the presenting symptom predicted an lengthened total diagnostic delay. Diagnostic delay had no impact on the tumour size at time of diagnosis or on the pre- and post-treatment morbidity.

Conclusions: Delays in the diagnosis of vestibular schwannoma have shortened since the introduction of magnetic resonance imaging. Longer diagnostic delays do not seem to have significant consequences.

Key words: Acoustic Neuroma; Magnetic Resonance Imaging; Diagnosis

Introduction

Little is known about diagnostic delays in vestibular schwannoma. In two reports by Thomsen and Tos,[1,2] vestibular schwannoma diagnosis was delayed in 78 per cent of patients for more than one year from the onset of symptoms; the mean delay for these patients was seven years. Van Leeuwen *et al.*[3] and Traquina *et al.*[4] reported that the average vestibular schwannoma diagnostic delay was approximately four years. In these four studies, data on symptoms and diagnostic delays were drawn from retrospective questionnaires[1,2] or retrospective chart reviews.[3,4] This approach can lead to bias, since the data concerning the onset and duration of symptoms are created after the patient is aware of the diagnosis of the intracranial tumour. The impact of diagnostic delay was evaluated by van Leeuwen *et al.*, who found that patients with a longer diagnostic delay had larger tumours, although this trend was not statistically significant.[3]

These four studies used patient data originating in the 1970s and 1980s, when vestibular schwannoma diagnosis was based on audiological studies, especially brainstem auditory evoked potentials, and computed tomography (CT).[5] Today, enhanced magnetic resonance imaging (MRI) is the recognised 'gold standard' for vestibular schwannoma screening, because of its superior sensitivity and specificity[6,7] and cost-effectiveness.[8] The introduction of MRI screening had been expected to shorten the delay between symptom onset and vestibular schwannoma diagnosis.[3] The aim of the present study was to characterise the current delay in vestibular schwannoma diagnosis, and to evaluate its impact on tumour size and symptoms at the time of diagnosis, as well as on symptoms or disabilities after treatment, in a relatively recent patient series from the era of routine MRI investigation for vestibular schwannoma. Primary healthcare data completed at the time of initial medical visit (assumed to be more accurate than retrospective recall) were used to assess the onset of presenting symptoms and the diagnostic delay.

Patients and methods

We identified from the hospital registers all consecutive patients diagnosed on MRI as having a schwannoma of the VIIIth cranial nerve (vestibular schwannoma; International Classification of Diseases

From the Department of Otorhinolaryngology, Kanta-Häme Central Hospital, Hämeenlinna, and the *Department of Otorhinolaryngology, Oulu University Hospital, Finland.
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code D33.3), within the Oulu University Hospital district (northern Finland; population approximately 700 000) between 1998 and 2006 and within the Kanta-Häme Central Hospital district (the district is located within south-central Finland; population approximately 170 000) between 1992 and 2006.

Detailed data on the first medical visit and the onset of the presenting symptom were collected from the primary care patient charts completed at presentation, before the true nature of the illness had been revealed. The presenting symptom was defined as the symptom which primarily resulted in the medical consultation ultimately leading to vestibular schwannoma diagnosis.

We collected the following clinical patient and tumour data from the hospital records: date of MRI diagnosis; symptoms and hearing level at time of diagnosis; size and location of tumour (i.e. intracanalicular versus extracanalicular or cerebellopontine); and selected treatment regimen (watchful waiting or microsurgery). For patients treated actively with microsurgery ($n = 49$), we also collected data on symptoms and disabilities after primary treatment. Patient delay was defined as the time from the onset of the presenting symptom to the first primary care medical visit, and professional delay was defined as the time from that first medical consultation to MRI diagnosis. The total diagnostic delay equalled the patient delay plus the professional delay.

Statistical methods

For analysis of the impact of diagnostic delay on tumour size and clinical outcome, the delays were dichotomised using the following cut-off points: for patient delay, three months (median delay) and six months ($n = 35$ *vs* 24); for professional delay, four months (median delay) and one year ($n = 40$ *vs* 19); and for total delay, 14 months (median delay), two years ($n = 35$ *vs* 24) and five years ($n = 51$ *vs* 8). The statistical significance of the observed differences was tested with Fisher's exact test in cases of categorical data, and with the Mann–Whitney U-test (comparing medians) and with independent samples *t*-test (comparing means) in cases of continuous data.

Results

A total of 91 patients diagnosed with vestibular schwannoma during the study period were identified (51 from Oulu University Hospital and 40 from Kanta-Häme Central Hospital). Nine patients (10 per cent) were diagnosed incidentally due to MRI or CT performed for another ailment; these patients were excluded from the delay analysis. Of the remaining 82 patients, 59 (72 per cent) had sufficient primary healthcare data from the time of symptom onset to allow analysis of diagnostic delay. The patient population included in the delay analysis did not differ from the whole study population in terms of age (mean age 53 *vs* 53 years, respectively), sex (males 46 vs. 47 per cent), tumour location (intracanalicular 34 per cent *vs* 33 per cent, respectively) or tumour diameter at the time of diagnosis (mean maximum diameter 1.8 *vs* 1.8 cm).

Table I presents the patient characteristics, presenting symptoms, tumour size and location, and primary treatment for all 91 patients. The patients' mean age was 53 years. 43 per cent were men. Unilateral hearing loss (45 per cent), tinnitus (18 per cent) and vertigo (8 per cent) were the most common presenting symptoms. Tumours were distributed equally

TABLE I

PATIENT, TUMOUR AND CLINICAL CHARACTERISTICS OF 91 CONSECUTIVE VESTIBULAR SCHWANNOMA PATIENTS DIAGNOSED IN FINLAND 1992–2006

Parameter	Intracanalicular tumours*	Extracanalicular tumours†	Total‡
Males (*n* (%))	13 (43)	30 (49)	43 (47)
Mean age (*n* (range); yrs)	54 (20–77)	52 (26–81)	53 (20–81)
Presenting symptom[a] (*n* (%))			
Unilateral HL	13 (43)	28 (46)	41 (45)
Tinnitus	5 (17)	11 (18)	16 (18)
Vertigo	4 (13)	3 (5)	7 (8)
Headache/pain	–	2 (3)	2 (2)
Ear fullness	–	1 (2)	1 (1)
Facial weakness	–	2 (3)	2 (2)
Other[b]	–	4 (7)	4 (4)
None[c]	4 (13)	5 (8)	9 (10)
Data missing	4 (13)	5 (8)	9 (10)
Symptoms at diagnosis (mean *n* (range))	1.4 (0–3)	1.6 (0–5)	1.4 (0–5)
Max tumour diameter on MRI (mean cm (range))	1.4 (0.3–3.0)	2.1 (0.8–5.5)	1.8 (0.3–5.5)
Primary treatment (*n* (%))			
Watchful waiting	17 (57)	25 (41)	42 (46)
Microsurgery	13 (43)[d]	36 (59)[e]	49 (54)
Stereotactic surgery	–	–	–

*$n = 30$ (33%); †$n = 61$ (67%); ‡$n = 91$. [a]Symptom that prompted first consultation ultimately leading to diagnosis. [b]Acute neurological deterioration ($n = 1$); sensory loss of trigeminal nerve ($n = 3$). [c]Incidental diagnosis (magnetic resonance imaging (MRI) or computed tomography performed for another ailment). [d]Retrosigmoid (suboccipital) approach, $n = 6$ (46%); translabyrinthine approach, $n = 5$ (38%); middle fossa approach, $n = 2$ (15%). [e]Retrosigmoid (suboccipital) approach, $n = 18$ (50%); middle fossa approach, $n = 16$ (44%); data missing, $n = 2$ (6%). Yrs = years; HL = hearing loss; max = maximum

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between the right (48 per cent) and left (52 per cent) sides. The mean maximum tumour diameter on MRI was 1.4 cm for intracanalicular tumours and 2.1 cm for extracanalicular tumours. During the study period, stereotactic surgery was not available in Finland; thus, all patients receiving active treatment (54 per cent) underwent open microsurgery.

Of the 59 patients included in the delay analysis, we obtained median values of three months for patient delay, four months for professional delay and 14 months for total delay (Table II). The total delay was longer than one year in 31 patients (52 per cent). The median total delay was not significantly affected by sex (17 *vs* 12 months for men and women, respectively; $p = 0.60$), age (16 *vs* 13 months for patients younger and older than 65 years, respectively; $p = 0.61$) or tumour location (24 *vs* 12 months for intracanalicular and extracanalicular tumours, respectively; $p = 0.36$). On the other hand, median total delay was longer among patients with unilateral hearing loss as the presenting symptom (24 months, versus nine months among other patients; $p = 0.042$). Exceptionally long delays were relatively rare; patient delay was over two years in six patients (10 per cent), professional delay was over five years in seven patients (12 per cent), and total delay was over five years in eight patients (14 per cent). The length of patient, professional and total diagnostic delays remained unchanged over the duration of the study period (comparing patients diagnosed in 1992–1999 and in 2000–2006; data not shown).

The diagnostic delays, when dichotomised, had no impact on the maximum tumour diameter at the time of diagnosis or on the pre- or post-treatment morbidity (Table III). The differences were equally insignificant, both clinically and statistically, with all the other cut-off points studied. Moreover, the diagnostic delays had no impact on tumour size at the time of diagnosis or on pre- or post-treatment morbidity, even when analysed separately for intra- and extracanalicular tumours.

TABLE II

DIAGNOSTIC DELAYS IN 59 VESTIBULAR SCHWANNOMA PATIENTS WITH DATA AVAILABLE, DIAGNOSED IN FINLAND 1992–2006

Delay (mths)	Intracanalicular tumours*	Extracanalicular tumours†	Total‡
*Patient***			
Mean	14	8	10
Median	3	2	3
Range	0–66	0–60	0–66
Professional§			
Mean	24	22	23
Median	5	5	4
Range	0–192	0–222	0–222
Total			
Mean	38	29	33
Median	24	12	14
Range	1–193	0–234	0–234

*$n = 20$ (34%); †$n = 39$ (66%); ‡$n = 59$. **From onset of presenting symptom to first medical visit. §From first medical visit to magnetic resonance imaging diagnosis. Mths = months

Facial nerve paresis at the time of diagnosis predicted larger tumours (mean tumour diameters of 3.2 *vs* 1.8 cm for patients with and without facial weakness, respectively; $p = 0.013$). Also, post-treatment facial paresis was significantly associated with tumour size ($p = 0.008$). None of the other individual pre- or post-treatment symptoms had a significant correlation with tumour size (data not shown). However, among patients with four or more presenting symptoms, the mean tumour diameter was 3.3 cm, versus 1.8 cm among the rest of the patients ($p = 0.015$).

Discussion

The delays in vestibular schwannoma diagnosis observed in this study were surprisingly short, and were markedly shortened compared with results from the 1970s and 1980s.[1–4] This is most probably due to the routine use of MRI, it is fast and its results are unambiguous and definite.

The only significant determinant of lengthened delay was unilateral hearing loss as the presenting symptom; this is understandable, since unilateral hearing deficiency is more likely to be considered by the patient and by the primary care physician as a less threatening ailment than tinnitus, vertigo, pain or facial weakness.

The length of diagnostic delay had no impact on the tumour size at time of diagnosis or on the pre- or post-treatment morbidity. Previously, a longer delay had been thought to automatically result in a larger tumour at the time of diagnosis,[2] even though data supporting this view were far from conclusive.[3] This finding could be explained by variation in the growth behaviour of schwannomas; aggressive tumours might grow and cause symptoms more rapidly, resulting in a shorter delay in the patient's decision to seek professional help and in the physician referring the patient for further studies. In addition, the fact that our patients' diagnostic delays were shorter compared with earlier research may partly explain why these delays did not significantly affect the tumour size at the time of diagnosis.

Determining the duration of delays in vestibular schwannoma diagnosis can be difficult. The timing of the onset of the first symptoms is very unlikely to be accurate; the patients are often elderly and may have had impaired hearing for years, regardless of the tumour. Also, symptoms such as headache, tinnitus and vertigo are commonplace among healthy people. Recollection of the onset and duration of first symptoms is likely to be biased, especially after the discovery of an intracranial tumour, and this limitation is shared by the previously published studies on the subject.[1–4] Also, these previous studies, with one exception,[3] did not evaluate the impact of diagnostic delay on clinical parameters.

Moreover, these previous studies dated from before the era of routine MRI investigation of vestibular schwannoma. Such imaging has completely changed vestibular schwannoma diagnostics. The present study covered a much more recent period

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TABLE III

IMPACT OF DIAGNOSTIC DELAYS ON MRI TUMOUR DIAMETER AND ON PRE- AND POST-TREATMENT MORBIDITY, IN VESTIBULAR SCHWANNOMA PATIENTS WITH DATA AVAILABLE, FINLAND 1992–2006

Parameter	Patient delay			Professional delay			Total delay		
	<6 mth*	≥6 mth†	p[a]	<1 yr‡	≥1 yr**	p[a]	<2 yr§	≥2 yr#	p[a]
Max tumour diameter on MRI (cm)									
Mean	1.9	1.8	0.66	1.9	1.8	0.72	1.9	1.8	0.79
Median	1.6	1.8	0.65	1.8	1.4	0.31	1.8	1.6	0.60
Range	(0.8–4.2)	(0.3–4.0)		(0.3–4.0)	(0.5–4.2)		(0.3–4.0)	(0.3–4.2)	
Pre-treatment morbidity (n (%))									
Hearing loss[b]	22 (63)	13 (54)	0.50	24 (60)	11 (58)	0.60	21 (60)	14 (58)	0.58
Tinnitus	19 (54)	12 (50)	0.43	21 (53)	10 (53)	0.53	19 (54)	12 (50)	0.43
Vertigo	13 (37)	9 (38)	0.55	15 (38)	7 (37)	0.54	13 (37)	9 (38)	0.55
Post-treatment morbidity[c] (n (%))									
Deafness[d]	23 (92)	16 (94)	0.60	25 (93)	14 (93)	1.0	20 (91)	19 (95)	0.53
Tinnitus	2 (8)	3 (18)	0.33	4 (15)	1 (7)	0.40	3 (14)	2 (10)	0.53
Vertigo	9 (36)	4 (24)	0.29	10 (37)	3 (20)	0.21	9 (41)	4 (20)	0.11
Headache/pain	4 (16)	3 (18)	0.62	4 (15)	3 (20)	0.49	4 (18)	3 (15)	0.53
Facial weakness	11 (44)	11 (65)	0.17	15 (56)	7 (47)	0.39	13 (59)	9 (45)	0.21

Patient delay = from presenting symptom onset to first medical visit; professional delay = from first medical visit to radiological diagnosis of vestibular schwannoma. *n=35; †n = 24; ‡n = 40; **n = 19; §n = 35; #n = 24. [a]Fisher's exact test in cases of categorical data, Mann–Whitney U-test (comparing medians) and independent samples t-test (comparing means) in cases of continuous data. [b]Pure tone average at 0.5, 1 and 2 kHz ≥ 30 dB in the affected ear. [c]Permanent morbidity; only patients with active primary treatment (surgery) included (n = 42). [d]Non-serviceable hearing even with a hearing device; data missing for one patient. Mth = months; max = maximum; MRI = magnetic resonance imaging

(1992–2006), with MRI screening in use the whole time.

- **The existing literature on diagnostic delays in vestibular schwannoma treatment is from the 1970s and 1980s, before routine use of magnetic resonance imaging**
- **A longer diagnostic delay had been thought to result in larger tumours at the time of diagnosis, although supporting data were not conclusive**
- **This study evaluated delays in vestibular schwannoma diagnosis and their impact on tumour size and on pre- and post-treatment morbidity, using relatively recent patient data**
- **Diagnostic delays have shortened markedly, and longer diagnostic delays do not appear to significantly affect tumour size or patient morbidity**

The data on first symptoms and the consequent first medical consultation were obtained from primary healthcare records completed at first presentation, before the true nature of the ailment had been discovered. While a prospective study evaluating delays in tumour diagnosis is virtually impossible to conduct, the method employed in the present study minimised the possibility of methodological bias with respect to the timing of symptom onset.

Patient delay can be prolonged because the symptom (e.g. slight, unilateral hearing loss) does not seem urgent or dangerous, or, in cases of more dire symptoms, because of conscious denial.[9] In the case of healthcare professionals, the reasons behind longer delay are more straightforward. The symptoms can often be considered mundane, especially when compared with the rarity of vestibular schwannoma. Also, screening tests used to indicate the need for MRI can be misleading; audiometry and calorimetric testing of vestibular function can be normal, especially in the case of small tumours, and testing for brainstem auditory evoked potentials may be unreliable when hearing loss is profound.[10]

Even though efforts could be made to shorten professional delay in vestibular schwannoma diagnosis, by better training of primary care physicians (e.g. by emphasising the possibility of vestibular schwannoma as a cause of unilateral hearing loss or tinnitus), the results of this study do not mandate this. A longer diagnostic delay does not seem to have serious implications, and it is questionable whether vestibular schwannoma patients benefit or suffer from earlier diagnosis; good results gained with a 'watchful waiting' approach[11–13] suggest that a considerable portion of such tumours remain small for a long time and do not require active treatment. Among patients with small tumours, early diagnosis of vestibular schwannoma, with possible consequent surgery, may well represent a worse option than delayed diagnosis, in terms of future life quality.

Conclusion

Delays in vestibular schwannoma diagnosis have shortened markedly with the introduction of MRI. In this study, prolonged diagnostic delay did not seem to have significant consequences in terms of tumour size, symptoms at the time of diagnosis or post-treatment morbidity.

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Address for correspondence:
Dr Heikki Teppo,
Department of Otorhinolaryngology,
Kanta-Häme Central Hospital,
FIN-13530 Hämeenlinna, Finland.

Fax: +358 3 6292933
E-mail: heikki.teppo@khshp.fi

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