The Journal of Laryngology & Otology
December 2000, Vol. **114,** pp. 935–939

Cystic vestibular schwannoma: surgical outcome

Petra Fundová, M.D.*†, Samih Charabi*, D.M.Sc., Mirko Tos, D.M.Sc.*,
Jens Thomsen, D.M.Sc., F.R.C.P.S. (Glas.)*

Abstract
We investigated the proportion of the cystic form of vestibular schwannoma and assessed the results of surgery in this subtype of the condition. The definition of cystic vestibular schwannomas was based on the following criteria: per-operative identification of cystic components; occurrence of the hypodense/hypointense areas on computed tomography (CT) and/or magnetic resonance (MR); and histological verification of S-100 protein membrane-like structures. In a study of 773 Danish patients with vestibular schwannomas, 44 (5.7 per cent) displayed cystic components. The outcome of surgery on 44 cystic vestibular schwannoma (mean tumour size 39 mm) was evaluated and compared with that for 151 solid grant vestibular schwannoma (mean tumour size 49.8 mm). Per-operatively, we found a substantially higher adherence to different intracranial structures in the solid giant vestibular schwannoma compared with the cystic vestibular (95 per cent *vs* 70 per cent for brainstem, 91 per cent *vs* 59 per cent for trigeminal nerve, 85 per cent *vs* 45 per cent for cranial nerves X and XI, 67 per cent *vs* 32 per cent for dura). Nevertheless, the preservation of the facial nerve function was much better in patients with solid giant vestibular schwannoma compared with those with cystic vestibular schwannoma (House-Brackmann facial nerve dysfunction grade 6 (one year post-operative): 27 per cent *vs* 41 per cent, respectively $p<0.04$). We conclude that the cystic components in vestibular schwannoma are associated with a less favourable surgical outcome, probably due to the rapid tumour growth and symptoms caused by compression of the posterior fossa structures.

Key words: Neuroma, Acoustic; Treatment Outcome

Introduction

Vestibular schwannoma is one of the most common intracranial tumours, which arise from the Schwann cells of the nerve sheath. Histologically the tumour is usually composed of Antoni type A and B tissue. Type A represents a more compact tissue, with elongated spindle cells, in irregular streams and with a tendency to palisading. Type B tissue, often intermingled with type A, is characterized by a loose texture, often with sponginess and cyst formation.[1]

Cystic vestibular schwannomas are observed routinely in radiology.[2,3] A few reports described the histological characteristics of these tumours. Cystic formation in schwannomas was initially thought to be related to degenerative changes or coalescence of microcysts in Antoni B tissue.[4] However, recently it has been shown that the cystic tumours contain an enlargement of the B tissue type, which is surrounded by a membrane-like structure composed of A cell type.[5]

The incidence of cystic formation in the vestibular schwannomas varies from 11.3 per cent[6] to 24 per cent[2] and 48 per cent.[3] However, in the latter studies cystic tumours were defined only according to the neuroradiological imaging. In the study of Charabi *et al.*[7] three criteria were required to be present before terming a tumour as cystic: the presence of a hypodense/hypointense area on CT/MR, per-operative identification of the cystic elements, and histological verification of the presence of S-100 positive membrane.[7]

The cystic nature of vestibular schwannoma is believed to be associated with faster tumour growth, shorter symptom duration, and with facial nerve involvement.[7,8]

The aim of this study was to investigate the incidence of cystic vestibular schwannomas and to assess the results of surgery in these types of vestibular schwannomas. In addition, it was evaluated whether the presence of cystic components in vestibular schwannoma may be relevant and important for predicting the post-surgical outcome.

From the Department of Otolaryngology – Head and Neck Surgery*, Gentofte University Hospital, Copenhagen, Denmark and the Department of Otorhinolaryngology†, 3rd Medical Faculty, Charles University, Prague, Czech Republic.
Accepted for publication: 10 July 2000.

https://doi.org/10.1258/0022215001904653 Published online by Cambridge University Press

Materials and methods

The reports of 773 patients with vestibular schwannoma were retrospectively reviewed. Two groups of tumours were separated: a group of 44 cystic vestibular schwannomas and a group of 151 solid giant vestibular schwannoma. The cohort of 44 tumours was separated as cystic tumours according to the following three criteria: the surgical protocols in all these 44 patients showed significant cystic elements, the imaging studies in these patients demonstrated CT- or MR-evidence of cyst formation, and the immunohistochemical staining revealed positivity for S100 protein in all of these 44 patients. None of the other 729 solid vestibular schwannomas exhibited any of these three characteristics.

There were 23 women (52.3 per cent) and 21 men whose ages ranged from 23 to 77 years (mean 52.7 years). Their symptoms developed mostly within less than four years, with the exception of 10 cases in which four patients have complained of hearing deterioration over the period of five to 10 years and six over 10 years. The clinical history ranged from six months to 27 years (mean 4.2 years). In 30 cases the initial symptom that led the patients to consult their physician was a slowly progressing hearing impairment. Seven patients suffered from vertigo as a primary symptom. Two patients had headache, and five of the patients had tinnitus. Pre-operatively, 17 patients had symptoms from the Vth (13 patients) and/or VIIth cranial nerve (five patients), which included facial nerve palsy (House-Brackmann grade 2 (HB2) in one case, HB3 in three cases, and HB4 in one case), facial and/or corneal dysaesthesia and/or paraesthesia. Twelve patients had symptoms of increased intracranial pressure with radiologically verified obstructive hydrocephalus. Before the surgery none of the cystic tumours had had previous radiotherapy.

The maximum extrameatal extension of the cystic tumours varied between 10 to 60 mm (mean 39 mm). There were five tumours classified as medium (10–25 mm), 14 tumours as large (26–40 mm), and 25 tumours were classified as giant tumours (>40 mm). The size data relates to distances of extrameatal expansion.

In this study we compared the surgical outcome of 44 cystic vestibular schwannoma with a group of 151 solid giant tumours, which represents the most at risk group within the non-cystic vestibular schwannoma. These solid giant tumours were selected according their size with extrameatal diameter of more than 40 mm. They did not exhibit any of the three stated criteria for cystic tumours.

None of the patients in both groups was a candidate for hearing preservation surgery with a hearing loss of 30 dB or less than 30 dB and a speech discrimination of 70 per cent or better.

The significance of differences was elevated using Student's *t*-test and $p<0.05$ was considered significant.

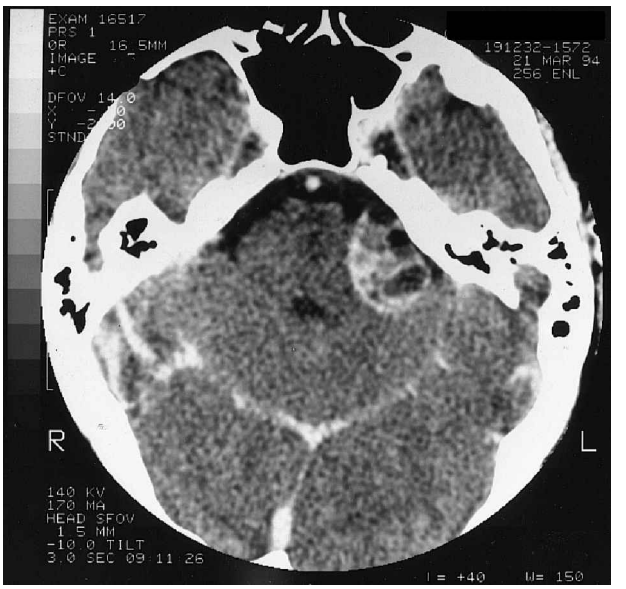

FIG. 1

Enhanced axial computed tomography image reveals the intratumoral cystic elements.

Results

In the period from the beginning of 1976 to October 1996 773 patients were operated on. Forty-four tumours (5.7 per cent) were termed to be cystic according to the following criteria: the per-operative identification of cystic elements, the occurrence of the hypodense/hypointense areas on CT or MR (Figures 1 and 2), and the histological verification of an S-100 positive membrane-like structure. We observed that the duration of the symptoms tended to be short, and it often involved functional impairment of the facial nerve as compared with their counterparts with solid giant tumours.

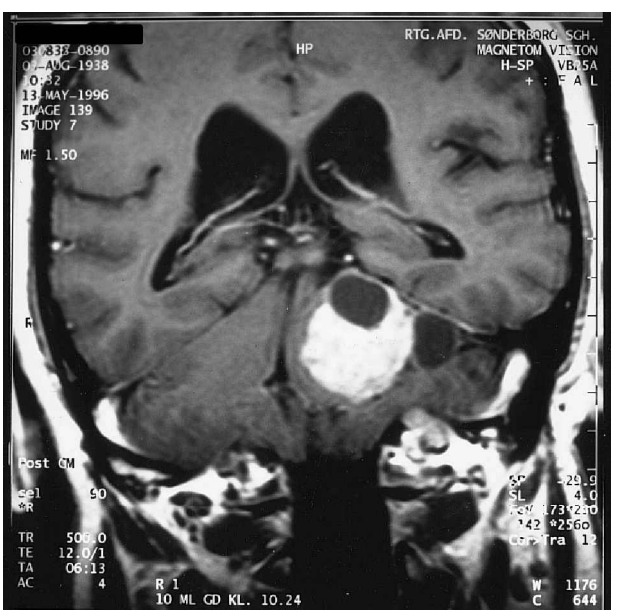

FIG. 2

Axial enhanced T1 magnetic resonance imaging demonstrates intratumoral hypointense areas.

https://doi.org/10.1258/0022215001904653 Published online by Cambridge University Press

TABLE I
CYSTIC VESTIBULAR SCHWANNOMA (VS) VERSUS SOLID GIANT VS: ADHERENCES TO DIFFERENT INTRACRANIAL STRUCTURES

	Cystic VS (n = 44)	Solid giant VS (n = 151)
Brainstem	70%	95%
Trigeminal nerve	59%	91%
Cranial nerves X and XI	45%	85%
Dura	32%	67%

Peroperative findings

In 70 per cent the cystic vestibular schwannomas adhered to the brainstem, in 59 per cent to the trigeminal nerve, in 45 per cent to the Xth and XIth cranial nerve, and in 32 per cent to the dura. The adherence of solid giant tumours to above mentioned intracranial structures is substantially higher as shown in Table I (95 per cent, 91 per cent, 85 per cent, 67 per cent, respectively).

Post-operative facial nerve function

At surgery the facial nerve was anatomically intact in 68 per cent of cystic vestibular schwannoma versus 79 per cent solid giant tumours (Table I). The post-operative facial nerve functions was evaluated according to the House-Brackmann classification system. Post-operatively (Table III), 12 (27 per cent) patients with cystic vestibular schwannoma and 47 (31 per cent) patients with solid giant tumour had a normal facial nerve function (HB1). In cystic vestibular schwannoma one patient (two per cent), two patients (five per cent) and six patients (14 per cent) classified as HB2, HB3 and HB4, respectively. In the solid giant tumour group 12 patients (eight per cent), 17 patients (11 per cent), and 19 patients (13 per cent) were classified as HB2, HB3, and HB4, respectively.

However, it should be stressed that substantially more patients (n = 18, 41 per cent) with cystic vestibular schwannoma displayed a complete loss of function of the facial nerve (HB6) compared to 41 (27 per cent) with solid giant tumour. The difference was statistically significant at level $p<0.04$ (Table III).

Complications

The tumour removal in cystic vestibular schwannoma and in solid giant tumour was complete in 39 (89.6 per cent) and 124 (82 per cent) patients, respectively (Table IV). Deliberate subtotal tumour removal in cystic vestibular schwannoma was performed in five (11.4 per cent) patients for various reasons. These included technical problems and the risk of bleeding, brainstem involvement and oedema of the cerebellum (Tables I and IV). One patient died six days after surgery due to a lesion in the brainstem (Table V). In clinical follow-up one year after surgery no recurrence was found in 39 patients with complete removal of tumour. In two patients the remaining tumour could easily be removed via the translabyrinthine route three and six months later after primary operation. In two patients the residual tumour is *in situ*. The total percentage of all listed complications (Table V) was substantially higher in patients with cystic vestibular schwannoma (31 per cent) compared to the group with solid tumours (20.5 per cent), $p<0.06$.

TABLE II
CYSTIC VESTIBULAR SCHWANNOMA (VS) VERSUS SOLID GIANT VS: PERI-OPERATIVE PRESERVATION OF FACIAL NERVE

	Cystic VS (n = 44)	Solid giant VS (n = 151)
Intact	68%	79%
Thin	–	10%
Not preserved	32%	11%

TABLE III
CYSTIC VESTIBULAR SCHWANNOMA (VS) VERSUS SOLID GIANT VS: POST-OPERATIVE FACIAL NERVE DYSFUNCTION AFTER ONE YEAR [HOUSE-BRACKMANN (HB) CLASSIFICATION]

	Cystic VS (n = 44)	Solid giant VS (n = 151)	p
HB 1	27%	31%	0.31
HB 2	2%	8%	0.09
HB 3	5%	11%	0.09
HB 4	14%	13%	0.43
HB 5	11%	10%	0.39
HB 6	41%	27%	0.04*

*Reconstructed facial nerve or VII–XII anastomoses are graded with HB 6.

Discussion

In previous studies based only on the neuroradiological appearance on CT or MR, without peroperative and immunohistological verification, the cystic vestibular schwannomas contributed by 9.7 per cent,[9] 11.3 per cent,[6] 20.5 per cent,[10] 24 per cent[2] and 48 per cent[3] respectively to all vestibular schwannomas. In this study we report that the incidence of cystic vestibular schwannoma is 5.7 per cent (Table VI). The discrepancy between the relatively lower incidence which was found by us and the relatively high incidence reported in previous studies could be explained by lower numbers of investigated tumours in previous studies. The largest group examined by Tali[6] consisted of 411 patients referred for clinical suspicion of vestibular schwannoma, the other groups consisted of less than 40 cases. Because of the large group of patients (n = 773) the incidence achieved in this study is probably more representative. In addition, in this study we used stricter criteria: the peri-operative identification of the cystic elements and the histological verification of the presence of S-100 positive membranes were employed beside the presence of hypodense/hypointense area on CT/MR. In the previous studies

TABLE IV
CYSTIC VESTIBULAR SCHWANNOMA (VS) VERSUS SOLID GIANT VS: SURGICAL RADICALITY

	Cystic VS (n = 44)	Solid giant VS (n = 151)
Total removal	89%	82%
Residual tumour	11%	18%

https://doi.org/10.1258/0022215001904653 Published online by Cambridge University Press

TABLE V
CYSTIC VESTIBULAR SCHWANNOMA (VS) VERSUS SOLID GIANT VS: MORTALITY AND COMPLICATIONS

Complication	Cystic VS ($n = 44$)	Solid giant VS ($n = 151$)
CSF leak	11%	9%
Haematoma	9%	3%
Cerebral oedema	4.5%	3%
Meningitis	4.5%	2%
Mortality	2%	4%
Total	31%	20.5% $p<0.06$

TABLE VI
PUBLISHED INCIDENCE OF CYSTIC VESTIBULAR SCHWANNOMAS

	All tumours	Cystic tumours
Kendall and Symon[9]	31	3 (9.7%)
Robbins and Marshall[10]	39	8 (20.5%)
Wallace *et al.*[2]	35	7 (20%)
Tali *et al.*[6]	80	15 (18.8%)
Charabi *et al.*[7]	571	23 (4%)
Jeng *et al.*[13]	27	13 (48%)
Pendl *et al.*[12]	148	9 (6.1%)
Current series	773	44 (5.7%)

by Wallace,[2] Jeng,[3] Tali,[6] Kendall,[9] Robbins[10] the cystic character of vestibular schwannoma was determined by neuroradiological imaging on CT and/or MR exclusively. Thus, the observed discrepancy in the frequency of cystic vestibular schwannoma could also be due to the fact that the different type of tissue in vestibular schwannoma could sometimes imitate cystic image on CT or MR scan; the number was higher because of a falsely positive cystic neuroradiological appearance.

In the last years much attention has been paid to the growth rate of vestibular schwannomas with and without cystic formation. Kameyama *et al.*[11] asserts that residual vestibular schwannomas with cyst formation showed rapid re-growth and required re-operation, as compared to residual vestibular schwannomas without cyst formation. Ki-67 staining revealed that the rapid volume increase in cystic vestibular schwannoma could be explained by an increase in the cyst volume rather, than by an increase in the growth rate of the tumour cells.[5] In the present study we have found that the occurrence of cystic vestibular schwannoma is usually connected with a substantially shorter period of symptom duration and with a substantially increased risk of accidental lesion of the facial nerve, compared with the group of solid vestibular schwannomas (Tables II and III). Thus, it could indirectly demonstrate faster growth potential in the group of cystic vestibular schwannoma.

The results of radiosurgery treatment in vestibular schwannomas with macrocystic components are quite unsatisfactory.[12] On the other hand, the results of surgical treatment in cystic vestibular schwannomas are better compared to radiosurgery treatment. Nevertheless as shown in this study, the outcome of the surgery in the cystic vestibular schwannoma group is still less favourable compared to the solid vestibular schwannomas.

It is thought that cystic formation in vestibular schwannoma could predict a more critical involvement of the neural tissue and a higher tendency toward bleeding also post-operatively.[13] It is believed that the cystic nature of vestibular schwannoma is connected with a rapid growth of the tumour, a poor surgical outcome and poor results of radiosurgery in this group of tumours as compared to the group of solid vestibular schwannomas.[14] Charabi *et al.*[7] asserts that the growth rate of cystic schwannomas is higher than that of solid tumour and that patients with cystic tumours are at risk of developing more symptoms, due ot the expansion of cystic elements, while the solid portion remains stationary.

Similar conclusions can be drawn from the findings in our large group of patients (n = 773) with vestibular schwannomas: those with cystic components (n = 44, 5.7 per cent) had more symptoms – development of hydrocephalus (n = 12; 27 per cent) and/or an accidental lesion of the facial nerve (Tables II and IV) than their counterparts. In accordance with Matthies *et al.*[13] we have found that the cystic nature of vestibular schwannoma is a critical finding decreasing the chances of functional nerve preservation at surgery (Tables III and IV). The cystic tumours differ from solid schwannomas by their rapid growth, a frequent involvement of facial nerve and by a lack of foreseeability of their biological behaviour.

In conclusion, in this study we report that cystic vestibular schwannomas occur less frequently than reported in previous studies based on neuroradiological imaging only. Surgical treatment of cystic vestibular schwannoma is favoured against radiosurgery[12] although cystic vestibular schwannoma still predicts worse surgical outcome compared to solid vestibular schwannomas. These should be taken into consideration when managing patients with cystic vestibular schwannomas. We propose that the finding of cystic vestibular schwannoma indicates a need for surgery as soon as possible since there is a possibility of the sudden expansion of the cystic elements, the involvement of facial nerve, and consequently a poorer surgical outcome.

References

1 Gruskin P, Carberry JN, Chandrasekhar SS. Pathology of acoustic tumors. In: House WF, Luetje CM, Doyle KJ, eds. *Acoustic Tumors. Diagnosis and Management*. 1st edn., San Diego: Singular Publishing Group Inc., 1997:27–85

2 Wallace CJ, Fong TC, Auer RN. Cystic intracranial schwannoma. *Can Assoc Radiol J* 1993;**44**:453–9

3 Jeng CM, Huang JS, Lee WY, Wang YC, Kung CH, Lau MK. Magnetic resonance imaging of acoustic schwannomas. *J Formos Med Assoc* 1995;**94**:487–93

4 Antoni NRE. Geschwulstgewebe, Geschwulstformen, Matrix. Histologie und Embryogenese der Neurofibrome. In; Antoni NRE, ed. *Über Rückenmarktumoren and Neurofibrome*. 1st edn., München: Kommissions-Verlag von J.F. Bergmann, 1920;311–8

5 Charabi S, Klinken L, Tos M, Thomsen J. Histopathology and growth pattern of cystic acoustic neuromas. *Laryngoscope* 1994;**104**:1348–52

https://doi.org/10.1258/0022215001904653 Published online by Cambridge University Press

6 Tali ET, Yuh WTC, Nguyen HD, Feng G, Koci TM, Jinkins JR, *et al.* Cystic acoustic schwannomas: MR characteristics. *Am J Neuroradol* 1993;**14**:1241–7
7 Charabi S, Tos M, Borgesen SE, Thomsen J. Cystic acoustic neuromas. *Arch Otolaryngol Head Neck Surg* 1994;**120**:1333–8
8 Lanser MJ, Jackler RK, Pitts LH. Intratumoral hemorrhage and cyst expansion as causes of acute neurological deterioration in acoustic neuroma patients. In: Tos M, Thomsen J, eds. *Acoustic Neuroma*. 1st edn. Amsterdam/ New York: Kugler Publications, 1992;229–34
9 Kendall B, Symon L. Investigation of patients presenting with cerebellopontine angle syndromes. *Neuroradiology***13**:65–84
10 Robbins B, Marshall WH, Jr. Computed tomography of acoustic neurinoma. *Radiology* 1978;**128**:367–70
11 Kameyama S, Tanaka R, Honda Y, Hasegawa A, Yamazaki H, Kawaguchi T. The long-term growth rate of residual acoustic neurinomas. *Acta Neurochir (Wien)* 1994;**129**:127–30
12 Pendl G, Ganz JC, Kitz K, Eustacchio S. Acoustic neurinomas with macrocysts treated with gamma knife radiosurgery. *Stereotact Funct Neurosurg* 1996;**66**(Suppl 1):103–11
13 Matthies C, Samii M, Krebs S. Management of vestibular schwannomas (acoustic neuromas): radiological features in 202 cases – their value for diagnosis and their predictive importance. *Neurosurgery* 1997;**40**:469–81
14 Kameyama S, Tanaka R, Kawaguchi T, Fukuda M, Oyanagi K. Cystic acoustic neurinomas: Studies of 14 cases. *Acta Neurochir (Wien)* 1996;**138**:695–9

Address for correspondence:
Professor Mirko Tos, M.D., D.M.Sc.,
Department of Otolaryngology - Head and Neck Surgery,
Gentofte University Hospital,
DK-2900 Hellerup,
Denmark.

Fax: 45 38777634

Petra Fundová, M.D. takes responsibility for the integrity of the content of the paper.
Competing interests: None declared
