Surgical Management of Endolymphatic Sac Tumors

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Abstract

Objective To review the cumulative experience of surgical management of endolymphatic sac tumors (ELSTs) in the literature. To review the experience with ELST management at a tertiary referral-based otology-neurotology practice and present our approach to management.

Study Design Retrospective patient review at a referral-based otology-neurotology practice.

Methods A review of all available records from the House Clinic of patients with ELSTs was performed. Presentation, diagnostic workup, microsurgical approach, and outcomes were reviewed. Literature review using MEDLINE of studies reporting surgical outcomes of patients with ELSTs was also performed.

Results Retrospective chart review revealed 18 patients with ELSTs who underwent surgical management. Grouping of these patients provides surgical outcomes on the largest group of mostly non-von Hippel-Lindau patients with ELSTs in the literature. Patients most commonly presented with sensorineural hearing loss, tinnitus, and dizziness. A majority of patients presented with large tumors exhibiting significant destruction of surrounding structures including the cochlea, vestibule, and internal auditory canal. Patients underwent microsurgical resection with the intent of complete resection.

Conclusions Although the extensive nature of ELSTs at time of diagnosis often precludes hearing preservation, complete microsurgical resection can be achieved safely with preservation of facial function in the majority of cases.

Keywords

- endolymphatic sac tumors
- facial nerve function
- hearing preservation

Introduction

Endolymphatic sac tumors (ELSTs) are rare, papillary adenomatous tumors that arise from the endothelium of the endolymphatic sac. These tumors were first recognized as a distinct pathologic entity with the report of Heffner in 1989.¹ These relatively rare but aggressive tumors cause local destruction of the temporal bone that can result in significant morbidity, including facial nerve weakness, hearing loss (HL), vestibular dysfunction, lower cranial nerve palsy, and occasionally death.² These tumors histologically are composed of

papillary and cystic components that infiltrate the surrounding bone.^{3,4} Radiologic imaging features include a retrolabyrinthine erosive lesion centered on the posterior petrous plate with intratumoral calcification on computed tomography and heterogeneous areas of increased signal intensity on T1- and T2-weighted magnetic resonance imaging (MRI) with variable enhancement using gadolinium (► **Fig. 1**).^{5,6}

Because of the rarity of these lesions, there are few series that document treatment outcomes. For this reason, we have summarized studies reporting on treatment outcomes in patients with ELSTs (► **Table 1**).^{1,7–13} We have previously

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Fig. 1 Radiographic findings of endolymphatic sac tumors. Axial temporal bone computed tomography (A). Axial T1-weighted magnetic resonance imaging (MRI; B). Axial T1-weighted MRI with gadolinium enhancement (C). Axial T2-weighted MRI (D).

reported on 14 patients with ELSTs.⁶ The surgical outcomes literature on ELSTs is reviewed and the outcomes of patients from this previous series, as well as new patients that have undergone surgical resection since this last report at the House Clinic, are presented. This represents the largest single-institution series of surgically treated patients with ELSTs with reported facial nerve and hearing preservation outcomes.

Methods

A literature review using MEDLINE was performed on studies reporting on outcomes of patients with ELSTs. Data collected when available included mean age at surgery, tumor size, number of patients with von Hippel-Lindau disease, percentage of patients with preservation of facial nerve function, percentage of patients who underwent surgical resection, complications of treatment, outcome status (no evidence of disease [NED], alive with disease [AWD], died of disease [DOD]), and mean follow-up in months with ranges.

A retrospective review of all available records from the House Clinic of patients with ELSTs was performed. Inclusion criteria was patients with a diagnosis of an ELST. Eighteen patients were identified, with 14 patients being reported on previously.⁶ Patients from this previously published report were included in this study.⁶ If charts of these patients were unavailable, outcomes were obtained from the published report.

Presentation, diagnostic workup, microsurgical approach, and outcomes were reviewed. More specifically, presenting signs and symptoms, preoperative diagnosis, surgical approach, complications of surgical treatment, pre- and post-operative facial nerve grade, and length of follow-up were collected. Determination of total resection at time of surgery, anatomical structures involved by the tumor, and presence of recurrent disease were recorded. Outcome status at most recent follow-up was designated as NED, AWD, or DOD. Hearing preservation was determined by preservation of preoperative hearing class (American Academy of Otolaryngology-Head and Neck Surgery [AAO-HNS] hearing classification). Facial function grade was recorded utilizing the House-Brackmann classification.

Results

A review of the records at the House Clinic identified 18 patients with ELSTs who underwent surgical resection between 1971 and 2011. Fourteen of the patients had been previously reported. When charts were available, the outcomes of these 14 patients were updated. Four additional patients were identified who underwent surgical resection of their tumors between 2001 and 2011. ▶**Fig. 2** shows the typical intraoperative appearance of these tumors.

▶**Table 2** presents the characteristics, symptoms, and physical findings of the patients on initial presentation. The study population consisted of 6 males and 12 females. The

Table 1 Literature review of outcomes in endolymphatic sac tumors

Study	Mean age at surgery (yrs)	Tumor size (cm)	VHL n (%)	Preservation of FN status n (%)	Surgical resection n (%)	Complications of treatment	Outcome	F/U mean (range) in mos
Pallanch et al, 1982 (<i>n</i> = 6)	50	N/A ^a	0 (0%)	N/A	4 (67%)	None	3 AWD 3 DOD	90 (1–180)
Heffner, 1989 (<i>n</i> = 16)	39	N/A ^b	0 (0%)	N/A	16 (100%)	Meningitis (1) Recurrence (1) Death (2)	11 NED 1 AWD 3 DOD 1 LTFU	61 (1–144)
Megerian et al, 1995 (<i>n</i> = 7)	52	N/A	1 (14%)	N/A	6 (86%)	Esophageal bleed after XRT leading to death Obstructive hydrocephalus	2 NED 4 AWD 1 DOD	46 (0.5–120)
Luff et al, 2002 (<i>n</i> = 3)	47	N/A	0 (0%)	N/A	3 (100%)	None	3 NED	60 (36–108)
Rodrigues et al, 2004 (<i>n</i> = 7)	52	N/A	0 (0%)	1 (14%)	7 (100%)	None	6 NED 1 AWD	69 (6–144)
Doherty et al, 2007 (<i>n</i> = 3)	35	5.1	0 (0%)	2 (67%)	3 (100%)	Pseudomeningocele Vocal cord palsy	3 NED	32 (12–48)
Bae et al, 2008 (<i>n</i> = 4)	48	3.8	0 (0%)	2 (50%)	4 (100%)	Iatrogenic FN injury with subsequent reanastomosis	3 NED 1 AWD	33 (14–61)
Timmer et al, 2011 (<i>n</i> = 9)	43	2.5	3 (33%)	7 (78%)	6 (67%)	Death of surgical complications (1)	5 NED 1 DOD	130 (37–252)
Current study (<i>n</i> = 18)	46	2.4	2 (11%)	10 (56%)	18 (100%)	HC requiring LPS HC requiring VPS CSF leak (3) LCN weakness (3) TN (1)	16 NED 1 AWD 1 DOD	67 (1–261)

Abbreviations: AWD, alive with disease; CSF, cerebrospinal fluid; DOD, died of disease; FN, facial nerve; F/U, follow-up; HC, hydrocephalus; LCN, lower cranial nerve; LPS, lumbar-peritoneal shunt; LTFU, lost to follow-up; N/A, data not available; NED, no evidence of disease; VHL, von Hippel-Lindau; VPS, ventriculoperitoneal shunt; TN, trigeminal neuralgia; XRT, radiotherapy.

^aTwo of six tumors had tumor sizes available (1.5 cm and 3 cm, respectively).

^bTwo of 16 tumors had tumor sizes available (4 cm and 6 cm, respectively).

mean age of the patients was 46 years. Mean tumor size was 2.4 cm, ranging from 1.0 to 3.3 cm. Sensorineural HL, tinnitus, and dizziness were common symptoms with 89%, 89%, and 72% of patients presenting with these symptoms, respectively. Four patients had symptoms characteristic of Meniere's syndrome and six patients presented with varying degrees of

facial weakness. Three patients exhibited preoperative cranial nerve (CN) V deficits and one patient exhibited lower CN involvement. Only two patients suffered from von Hippel-Lindau disease (patients 12 and 18).

As previously described, patient 7 was referred from an outside institution with a diagnosis other than ELST. This

Fig. 2 Intraoperative findings in patient 16. View depicts tumor arising from the area of the endolymphatic sac and duct, between the sigmoid sinus and posterior semicircular canal (left). Removal of tumor with resection of dura (right). mfd, middle fossa dura; ss, sigmoid sinus; *tumor.

Table 2 Patient characteristics and presenting signs and symptoms

Patient	Age at presentation (yrs)	Gender	Tumor size	SNHL	Tinnitus	Dizziness	Meniere's syndrome	Preoperative facial grade (HB)	Other cranial neuropathies	Preoperative diagnosis
1	57 61 65	F	N/A N/A N/A	Y	Y	Y	N	6 6 6	V3 V3 V3	Meningioma ELST ELST
2	43	F	N/A	Y	Y	Y	N	5	V3	Cholesteatoma
3	21	F	3.3	Y	Y	N	N	5	V3	Papillary adenoma
4	24	F	2.0	Y	Y	Y	Y	3	None	Meningioma
5	18	F	3.0	Y	Y	Y	Y	1	None	Hemangioma
6	45 48	M M	2.5 0.5	N	N	N	N	2 6	None None	ELST
7	50	M	1.0	Y	Y	Y	N	1	None	ELST-CG
8	54	F	5.5	Y	Y	N	N	6	IX	Paraganglioma (prior resections 1983, '84, '85, '86)
9	50	M	2.2	Y	Y	Y	N	1	None	Chondrosarcoma
10	33	M	3.1	Y	Y	Y	N	1	None	ELST
11	37	F	2.0	Y	Y	Y	Y	1	None	ELST
12	36	F	2.5	Y	Y	Y	N	1	None	ELST
13	55	F	4.3	Y	Y	Y	N	1	None	ELST
14	75	F	2.0	N	Y	Y	N	1	None	ELST
15	49	M	2.4	Y	Y	Y	N	1	None	ELST
16	45	F	3.4	Y	Y	Y	Y	1	None	ELST
17	72	F	1.0	Y	N	N	N	1	None	ME Adenoma
18	20	M	2.0	Y	Y	N	N	1	None	ELST
Total or Average ($\pm$ SD)	46 ($\pm$ 16)	6 M 12 F	2.4 ($\pm$ 1.0)	16/18 (89%)	16/18 (89%)	13/18 (72%)	4/18 (22%)	HB 1: 12/18 (67%) HB 2: 1/18 (6%) HB 3: 1/18 (6%) HB 5: 2/18 (11%) HB 6: 2/18 (11%)		

Abbreviations: CG, cholesterol granuloma; ELST, endolymphatic sac tumor; F, female; HB, House-Brackmann classification; M, male; ME, middle ear; N/A, not available; SD, standard deviation; SNHL, sensorineural hearing loss.

patient had undergone a transmastoid approach to drain a presumed cholesterol granuloma 2 years before being seen at our clinic. Patient 8 had undergone four prior resections of a presumed middle ear paraganglioma in 1983, 1984, 1985, and 1986. She later underwent external beam radiotherapy (5400 rads) for a large recurrence. Despite this, the tumor persisted and grew, extensively invading the jugular foramen and cavernous sinus, and encased the internal carotid.

All patients underwent microsurgical resection of their tumors. The surgical findings, approaches, and complications are noted in **Table 3**. A gross total removal was believed to be accomplished in all but the first surgery in the first patient. Patient 1 underwent multiple craniotomies with persistent growing disease despite resections that were believed to be gross total resections. In patient 6, a small recurrence, less than 0.5 cm, was noted 3 years after the initial surgery and a second retrolabyrinthine craniotomy was performed to remove this focus of recurrent tumor. Excluding these two patients, gross total resections were accomplished in the remainder of the patients with no recurrences.

A new onset of cranial neuropathies, excluding CN VII and VIII, in 4 (22%) patients constituted the most common postoperative complication. In one patient (patient 13), the CN IX paresis was temporary. Three (17%) patients developed a postoperative cerebrospinal fluid leak that responded to wound revision and lumbar drain. Two (11%) patients developed hydrocephalus requiring a ventriculoperitoneal shunt and a lumbar peritoneal shunt (LPS), respectively. The patient who underwent the LPS for hydrocephalus was a particularly obese female with an extensive 3.4 cm ELST. We suggest that this patient had existing comorbidities that predisposed the patient to this particular complication. There were no perioperative mortalities, infections, or thromboembolic complications.

The outcomes of surgery and follow-up are presented in **Table 4**. Follow-up ranged from 1 to 261 months with mean follow-up time of 67.0 months. Persistent or recurrent disease occurred in three patients. Two of these patients underwent multiple attempts at tumor removal, at our clinic (patient 1) or elsewhere (patient 8). Neither patient was diagnosed with ELST on initial pathologic examination. At last follow-up, patient 1 was noted to have a large, unresectable tumor. Patient 8 succumbed to her disease 3 years after her last surgery (17 years after the initial surgery). Both patients with persistent growing disease underwent external beam radiotherapy at some point in their treatment course. Patient 6 underwent a second retrolabyrinthine craniotomy to remove a small focus of recurrent or residual tumor and now remains without evidence of recurrent disease. None of the patients developed metastatic disease. Regarding the six patients with less than 1 year of follow-up, patients 7, 12, and 14 were lost to follow-up prior to achieving 1 year of follow-up. Patients 15, 16, and 18 are currently being followed. Gross total resection was achieved in all of these patients.

With respect to facial nerve function outcomes, all patients who presented initially with normal facial nerve function maintained normal facial nerve function with the exception of two patients (patients 5 and 16, House-Brackmann [HB]

grade II and V, respectively). Overall, 10 (56%) patients had a HB grade I and 11 (61%) patients exhibited a HB grade II or better. Two patients presented with normal hearing, and preoperative hearing class (AAO-HNS hearing classification) was preserved in these cases. Hearing preservation was attempted by retrolabyrinthine craniotomy in both cases. The tumor in patient 6 did not exhibit invasion of the vestibular labyrinth or the semicircular canals. Patient 14 required partial resection of the posterior semicircular canals followed by immediate sealing with bone wax.

Literature Review

In reviewing studies that reported treatment outcomes, we identified eight studies other than the current study reporting on outcomes of patients with ELSTs, some of whom underwent surgical resection (**Table 1**).⁷⁻¹³ Only about 100 cases have been reported in the past 100 years. Including the current study, a total of 73 patients with some information on outcomes has been reviewed. Of the patients with facial nerve outcomes ($n = 41$), 22 (53%) patients exhibited preservation of preoperative HB grade. Patients with von Hippel-Lindau disease constituted 8% ($n = 6$) of all the studied patients. Microsurgical resection was performed in 67 (92%) patients. Complications were experienced by 19 (26%) patients. With respect to most recent follow-up, 49 (67%) patients exhibited NED, 11 (15%) patients were AWD, 9 (12%) patients DOD, and 1 (1%) patient was lost to follow-up.

Discussion

ELSTs are low-grade papillary adenocarcinomas arising from the endolymphatic sac. These tumors are slow growing but can be locally aggressive. Previous publications have outlined our treatment bias, namely, a preference for complete surgical resection when possible, with earlier diagnosis and treatment associated with better functional outcomes. From our perspective, radiosurgical treatment of ELSTs remains controversial but may be utilized in cases where complete resection is impossible or in cases where tumor is unresectable.

In this study, we review 18 ELST patients who underwent surgical resection. This represents the largest single-institution series of surgically treated ELST patients. When compared with the published literature, our results compare favorably. Specifically, preoperative facial nerve grade preservation compared similarly to those published in the literature. Of the 12 patients who presented with an HB grade I preoperatively, 10 (83%) patients maintained their preoperative facial function. This particular outcome compares similarly to the recent study by Timmer and colleagues.¹³ Furthermore, 89% of patients show no evidence of disease at most recent follow-up. This compares similarly to rates published in the literature. Because it is a tertiary care clinic, patients often are referred in with extensive disease when it comes to ELSTs.

Of the two patients with normal hearing (AAO-HNS class A) who were referred that had tumors amenable to hearing preservation (no invasion of the vestibular labyrinth

Table 3 Operative approach, findings, and complications

Patient	Surgical approach	Total resection	Labyrinth	Jugular foramen	IAC	Mastoid facial nerve	ICA	Intracranial (beyond dura)	Complications
1	MFC (1971) TLC/TCC (1975) TLC/TCC (1979)	N Y ^a Y ^a	Y	Y	Y	N	Y	Y	CN IX, X, XI
2	TLC/TCC	Y	Y	Y	Y	N	Y	Y	CN IX, X, XI
3	TLC/ITFC/FNG	Y	Y	N	N	Y	N	N	None
4	TLC/FNG	Y	Y	N	N	Y	N	N	None
5	TLC	Y	Y	Y	N	N	N	N	CSF leak; TN
6	RLC (1997)	Y							
	RLC (2000)	Y	N	N	N	Y	N	N	Recurrence
7	TLC	Y	Y	N	N	N	N	N	None
8	ITFC type C; ICA and CN 7–11 sacrificed	Y	Y	Y	Y	Y	Y	Y	HC requiring VPS
9	TLC	Y	Y	N	Y	N	N	N	CSF leak
10	TLC, ITFC	Y	Y	N	Y	N	N	N	None
11	TLC	Y	Y	N	N	N	N	N	None
12	TLC	Y	Y	N	N	N	N	N	None
13	TLC, ITFC	Y	Y	Y	N	N	Y	N	Temporary CN IX; CSF leak requiring LD & wound revision
14	RLC/partial removal PSCC	Y	Y	N	N	N	N	N	None
15	TLC, blind sac closure EAC	Y	Y	N	Y	Y	N	N	None
16	TLC, ITFC	Y	Y	Y	Y	Y	N	Y	HC requiring LPS
17	TM	Y	N	N	N	N	N	N	None
18	TLC	Y	N	N	N	N	N	N	None
Total		17/18 (94%)	15/18 (83%)	6/18 (33%)	7/18 (39%)	6/18 (33%)	4/18 (22%)	4/18 (22%)	

Abbreviations: CN, cranial nerve; CSF, cerebrospinal fluid; EAC, external auditory canal; FNG, facial nerve graft; HC, hydrocephalus; IAC, internal auditory canal; ICA, internal carotid artery; ITFC, infratemporal fossa craniotomy; LD, lumbar drain; LPS, lumbar peritoneal shunt; MFC, middle fossa craniotomy; MI, myocardial infarct; PSCC, posterior semicircular canal; RLC, retrolabyrinthine craniotomy; TCC, transcochlear craniotomy; TLC, translabyrinthine craniotomy; TM, transmastoid; TN, trigeminal neuralgia; VPS, ventriculoperitoneal shunt.

^aThese were thought to be gross total resections; however, patient was noted to have persistent disease that was growing on radiologic monitoring.

Table 4 Patient follow-up and outcomes

Patient	Follow-up (mos)	Recurrence	XRT	Status	Facial grade (HB) (pre-postoperative)	Hearing preservation
1	261	Y $\times$ 3	Y (4800 Rad)	AWD	VI-VI	N
2	33	N	Y (7000 Rad)	NED	V-VI	N
3	105	N	N	NED	V-III	N
4	73	N	N	NED	III-IV	N
5	108	N	N	NED	I-II	N
6	88 54	Y N	Y N	AWD NED	II-VI VI-VI	Y Y
7	10	N	N	NED	I-I	N
8	18	Y	Y (5400 Rad)	DOD ^a	VI-VI	N
9	145	N	N	NED	I-I	N
10	152	N	N	NED	I-I	N
11	14	N	N	NED	I-I	N
12	1	N	N	NED	I-I	N
13	74	N	N	NED	I-I	N
14	6	N	N	NED	I-I	Y
15	5	N	N	NED	I-I	N
16	9	N	N	NED	I-V	N
17	115	N	N	NED	I-I	N
18	2	N	N	NED	I-I	N
Total ($\pm$ SD)	67.0 ($\pm$ 69.1)	3/18 (11%)	4/18 (22%)	NED: 16/18 (89%) AWD: 1/18 (6%) DOD: 1/18 (6%)		2/18 (11%)

Abbreviations: AWD, Alive with disease; DOD, died of disease; HB, House-Brackmann classification; NED, no evidence of disease; SD, standard deviation; XRT, radiotherapy. Note: Patients 7, 12, and 14 were lost to follow-up prior to achieving 1 year of follow-up. Patients 15, 16, and 18 are currently being followed.

^aPatient 8 died of her disease, developing aspiration and eventually dying from a myocardial infarction.

or semicircular canals), preoperative hearing class was preserved. Given the usually extensive nature of these tumors by the time of presentation and treatment, hearing preservation is not often possible. However, especially in patients with known von Hippel-Lindau disease, earlier diagnosis and complete surgical resection may make hearing preservation an option.^{14,15}

Given the extensive nature of the majority of the tumors treated in this series, the complication rates reported in this study are acceptable. Other cranial neuropathies, particularly of the lower cranial nerves, have been minimized in more recent patients. Overall, these outcomes are likely the result of both improved experience with ELST resection over time and earlier radiologic diagnosis and treatment with an emphasis on complete surgical resection.

The role of radiation in the treatment of ELSTs is controversial. In Heffner's original report, 11 of 12 tumors treated with total resection remained free of disease, whereas only 1 of 4 patients treated with subtotal resection followed by postoperative radiation therapy were free of disease.¹ Furthermore, of the three patients in our series treated with radiation, only one remained free of disease during the

follow-up period, with the other two patients either having persistent disease or dying as a result of disease progression. The utilization of chemotherapy in the treatment of ELSTs is even more controversial as no published cases exist of its use. In the authors' opinion, gross total resection is the treatment of choice for ELSTs and monitoring with serial MRI and subsequent re-resection would be favored over radiation or chemotherapy.

Limitations of this study include its retrospective nature and the wide range of follow-up, ranging from less than 1 year to approximately 22 years. Of these patients, three patients are currently being followed and three have been lost to follow up. We report the disease status at the end of the last follow-up. The experience with these rare tumors has been accumulated over a period of 40 years and has been retrospectively reviewed. Although freedom from disease cannot be assumed beyond the noted follow-up period, freedom from disease within the follow-up period is noted. Despite these limitations, this represents the largest series of surgically managed ELSTs from a single institution and does suggest that gross total resection of disease is an effective long-term treatment.

Conclusions

ELSTs are relatively slow growing but locally aggressive tumors for which we advocate complete surgical resection as the treatment of choice. Although the extensive nature of ELSTs at time of diagnosis often precludes hearing preservation, complete microsurgical resection can be achieved safely with preservation of facial function in the majority of cases.

Institutional Review Board

Institution: St Vincent Medical Center Institutional Review Board.

IRB Approval: Yes.

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