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The Gruppo Otologico experience of endolymphatic sac tumor

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Abstract

Objective: Endolymphatic sac tumor (ELST) is a rare low grade adenocarcinoma of the skull base. During the past decade the number of the reported cases has increased. This study exposes our experience in the management of ELST with a review of the literature.
Study design: Retrospective study of patients with ELST at a quaternary referral otology and skull base center.
Methods: A review of the records from the Gruppo Otologico revealed 7 patients treated for ELST. All papers containing series of three or more cases of ELST published in the English literature were selected for analysis.
Results: Hearing loss and tinnitus were present in almost all our cases. All of them were evaluated with audiometric tests, computed tomography and magnetic resonance imaging. All the patients were treated surgically with preservation of the facial nerve and preoperative embolization was performed in 5 patients. Genetic study was performed on all our cases and revealed the presence of von Hippel–Lindau syndrome in one patient who had the tumor as the initial manifestation of his syndrome. None of the patients received postoperative radiotherapy and one of them had recurrence of the tumor 13 years following surgery.
Conclusions: Complete surgical resection with preoperative embolization of large tumors is the mainstay treatment for ELST. The facial nerve should not be sacrificed unless it is totally invaded by the tumor. A long term follow up is recommended and the role of postoperative adjunctive radiotherapy is still controversial.

Keywords: Endolymphatic sac tumor; Adenoma of the endolymphatic sac; Adenocarcinoma of the endolymphatic sac; Heffner tumor; von Hippel–Lindau; Low grade papillary adenocarcinoma

1. Introduction

The endolymphatic sac is a neuroectodermal blind pouch that lies in the Trautmann's triangle midway between the internal auditory canal and the sigmoid sinus. The major role of the sac is the regulation of the endolymphatic fluid volume and pressure as well as its homeostasis and absorption of cellular debris [1].

Endolymphatic sac tumor (ELST) is a locally aggressive low grade adenocarcinoma that arises from the proximal rugose portion of the endolymphatic sac epithelium. It has been first described by Hassard et al. in 1984 as an adenoma of the endolymphatic sac that was found incidentally upon a decompression surgery of the sac in a patient presumed having Meniere's disease [2].

ELST is a rare sporadic entity, however it can be associated with von Hippel–Lindau (VHL) disease in 24% of the cases [3].

These tumors were misdiagnosed as paragangliomas, but with the recent advances in pathological and radiological

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Table 1
Patients demographic data and clinical findings.

Patient Nb	VHL	Age	Sex	Side	Tinnitus	Otalgia	Vertigo	Preop FN grading	Retrotympanic mass	Audiogram
1	N	33	F	R	Y	Y	N	1	Y	40 dB (SNHL)
2	N	20	F	L	Y	N	N	1	Y	60 dB (SNHL)
3	N	54	M	L	Y	N	Y	1	N	55 dB (mixed hearing loss)
4	N	23	F	R	Y	N	N	1	N	Dead ear
5	Y	25	F	R	N	N	N	3	Y	Dead ear
6	N	30	F	R	Y	N	Y	1	N	50 dB (SNHL)
7	N	33	M	R	Y	N	N	1	N	Dead ear

Abbreviations: F, female; L, left; M, male; N, no; R, right; Y, yes; Preop, preoperative; CN, cranial nerve; dB, decibel; FN, facial nerve; Nb, number; SNHL, sensorineural hearing loss; VHL, von Hippel–Lindau disease.

diagnostic tools, these tumors are more easily recognized and more reports are encountered in the literature.

In this manuscript we present the Gruppo Otologico experience with ELST in addition to a literature review and an analysis of all the previous reported series.

2. Materials and methods

After departmental and institutional review board approval, a retrospective chart review was conducted on all cases of ELST treated at the Gruppo Otologico medical center between January 1987 and December 2010.

Information collected included demographic data, tumor characteristics, audiometric tests, radiological exams, surgical treatment, adjunctive therapies and follow-up.

We performed also a detailed search in Pubmed and Medline database with a complete review of all the articles published in the English literature from 1984 until December 2010 using the following keywords: endolymphatic sac tumor, adenoma of the endolymphatic sac, adenocarcinoma of the endolymphatic sac, Heffner tumor, and von Hippel–Lindau. Publications containing series of three or more cases of ELST (including our series) were selected for analysis. We excluded two large studies done by Patel et al. and Mukherji et al. because they were focused only on radiological characteristics of ELST. A series reported by Kim et al. was abandoned because it included the same group of patients that was reported 3 years later by Lonser et al. [4].

3. Results

3.1. Patients

There were 7 patients, 2 men and 5 women. Mean age at presentation was 31.1 years with a range from 20 to 54 years. The most common presenting symptoms were hearing loss, tinnitus and vertigo. One patient had preoperative facial nerve weakness (House–Brackman grade III). None of our patients presented with lower cranial nerve deficit (Table 1).

All patients underwent pure-tone audiometry, imaging with computed tomographic (CT) scan and magnetic resonance imaging (MRI). All the patients had retrolabyrinthine location of the tumor with hyperintense focal signals on non-enhanced T1-weighted MRI with heterogeneous signal on T2-weighted MRI (Fig. 1). The size of the tumor ranged from 1 to 5 cm. Five patients underwent embolization.

All the patients underwent complete surgical resection of their tumors, the approach was chosen according to the tumor extension and the facial nerve was preserved in all the cases. None of the patients received postoperative radiotherapy. Genetic counselling was performed on all our cases and revealed the presence of VHL disease in only one patient. In this case the ELST was the initial manifestation of the disease (Table 2).

On histology, all tumors showed wide cystic cavities containing branching papillary projections with focal small glands (Fig. 2a and b). Some cysts exhibited more intensely eosinophil fluid and multiple vacuoles that closely simulate colloid follicles in the thyroid gland. In 2 cases, focal areas seemed to be more solid due to the coalescence of contiguous papillae and glands (Fig. 2a). Both cysts and papillae were lined by a monolayer of isomorphic cuboidal cells that progressively become flat and sometimes barely recognizable.

The axis of the papillae was hypocellular fibrous or oedematous with a capillary vascularization lying just below the epithelium. Occasional papillae were dilated and pseudocystic due to marked oedema of their stromal axis.

In all the specimens, recent and old stromal haemorrhages were present. In patients treated with preoperative embolization focal neutrophilic inflammation, necrosis and erosion were evident in the papillae.

Nuclei were homogeneous in size, mainly central or close to the luminal surface and presented finely granular chromatin with inconspicuous nucleoli. Cytoplasms were clear, optically empty or eosinophilic and faintly granular in some areas (Fig. 2c).

PAS positive diastase-sensitive heterogeneous granules were frequently detectable.

Immunohistochemistry played a major role in the diagnosis of ELST. In all the specimen examined the

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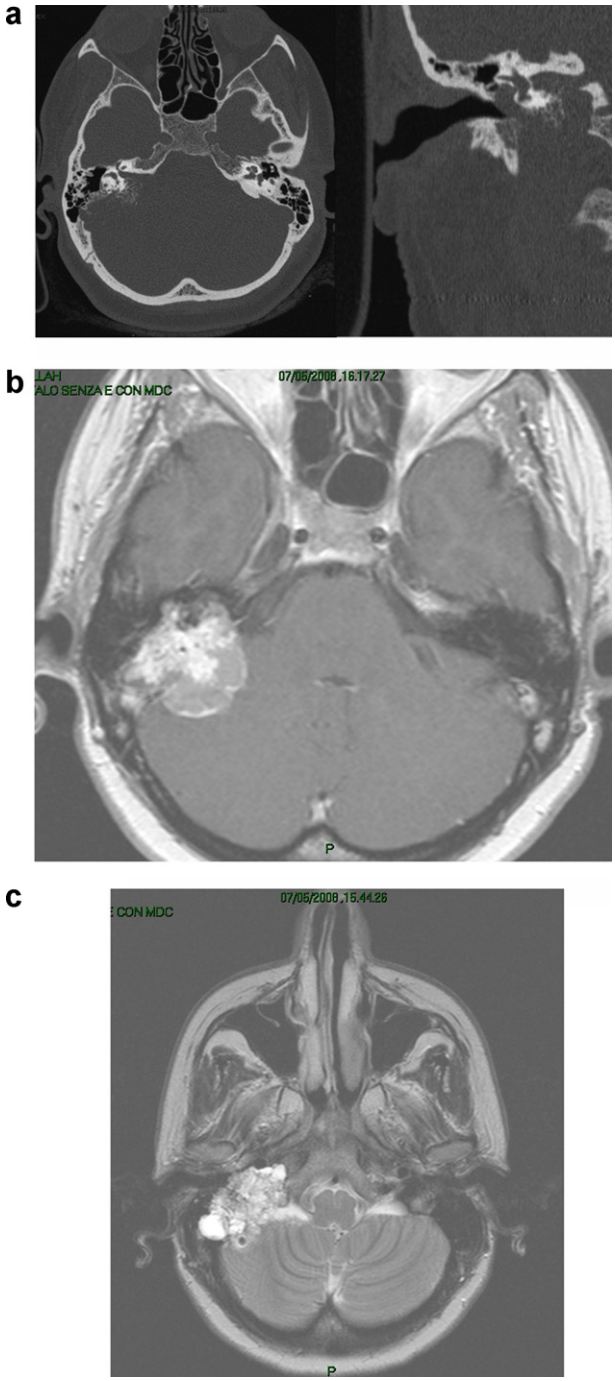

Fig. 1. (a) Axial and coronal CT scans showing a destructive mass epicentered on the right posterior petrous face of the temporal bone and erodes the internal auditory canal with central speculated calcifications. (b) T1 gadolinium-enhanced axial MRI scan demonstrated the heterogeneous enhancement of the tumor. (c) Axial T2-weighted MRI scan of an endolymphatic sac tumor demonstrating heterogeneous signal.

epithelial cells were positive for keratins (*clone AE1/AE3LP34*), vimentin (*clone V9*), Neuron Specific Enolase (NSE, *clone BBS/NC/VI-H1L*) which is highly specific for sac tissue (Fig. 2d and e). They were negative for Synaptophisin (*policlonal*) and in one patient few nervous cells were highlighted by S-100 in the axis of some large papillae. No mitosis has ever been detected. Proliferative activity recognized by MIB-1 antibody (*clone MM1*) was negligible (≤1%). Bone infiltration was evident in all studied specimens and in one case there was a tight connection between cystic structures and bone lamellae suggesting indolent bone remodeling (Fig. 2b).

Two patients developed post operative mild to moderate facial weakness (House–Brackmann Grade II, III). Only one patient had a transient unilateral vocal cord paresis.

On follow up, one patient had recurrence of the tumor 13 years following surgery discovered incidentally. The patient underwent a revision surgical intervention via a translabyrinthine approach and was free of disease at 7 years follow up clinically and radiologically.

Fifteen months after surgery the patient with VHL disease had new lesions on MRI in the cervical spine and left cerebellum. These lesions were resected at another center, pathology revealed the presence of hemangioblastoma. This patient remained free of disease for 8 years of follow up.

3.2. Literature review

Review of the publications in the English literature revealed 104 articles with 270 cases of ELST.

The majority of these articles are single case reports and only 16 publications fulfilled our criteria of inclusion for the analysis (3 or more cases/article).

Patient demographics, tumor characteristics, presenting signs and symptoms described in these articles (including our series) are summarized in Table 3.

There were 107 patients with female predominance (F/M: 3/2). Mean age at presentation was 38.1 years (15–75 years). VHL disease was found in 20.5% of the patients.

There was no site predilection for the tumor. The size ranged from 0.2 cm to 6.4 cm.

Hearing loss was the pertinent presenting symptoms (94%), followed by tinnitus (55%), vertigo (47%), facial palsy (32.7%), lower cranial nerve deficit (5%) and facial paresthesia (5%).

The interval between onset of symptoms and tumor diagnosis ranged from 2 to 276 months (median 67.5 months).

All the patients were treated surgically except one. The surgical approaches varied according to the hearing status and tumor stage. The surgical approaches are classified into two groups: hearing ablation surgeries (translabyrinthine, transchoclear, transotic or combined) used in 63% of the patients and hearing conservation surgeries (transmastoid, retrolabyrinthine transdural, suboccipital or combined) used in 35% of the cases.

Subtotal resection of the tumor was performed in 11% of the patients. Preoperative embolization has been used in 18.7% of the cases. Postoperative radiation therapy (fractionated or stereotactic) was given as an adjunctive therapy in 21.5% of the patients. One patient was treated primarily by radiation therapy and passed away shortly

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Table 2
Intraoperative findings and post operative outcomes.

Patient Nb	Tumor size	Embolization	App	Tumor extension							FN management	FN grade	Recurrence	Last F/U	Status
				LAB	ME	PCF	MCF Dura	JB	IAC	FN					
1	3.3 cm	Y	TLA	N	Y	Y	Mass effect	N	Y	N	–	1	N	30 m	NED
2	4 cm	Y	TO	Y	Y	Y	Mass effect	Y	Y	N	–	1	N	52 m	NED
3	3 cm	Y	TO + ITF A	Y	N	Y	N	Y	N	Y	Anterior rerouting	3	N	40 m	NED
4	3 cm	Y	TLA + ITF A	Y	N	Y	N	Y	Y	Y	Anterior rerouting	2	N	60 m	NED
5	5 cm	Y	TC	Y	Y	Y	Mass effect	N	Y	Y	Posterior rerouting	3	N	92 m	NED[a]
6	2 cm	N	TLA	Y	N	N	N	N	N	N	Skeletonization upon revision surgery	1	Y (13 years after surgery)	240 m	NED
7	1 cm	N	TLA	Y	N	N	N	N	N	N	–	1	N	72 m	NED

Abbreviations: m, month; N, no; Y, yes; App, approach; FN, facial nerve; F/U, follow up; IAC, internal auditory canal; ITF A, infra temporal fossa type A; JB, jugular bulb; LAB, labyrinth; ME, middle ear; MCF, middle cranial fossa; Nb, number; NED, no evidence of disease; PCF, posterior cranial fossa; TC, transcochlear, TO, trans-otic, TLA, translabyrinthine approach.

[a] But new appearance of a spinal cord, and L cerebellar hemangioblastoma 15 m after her surgery.

during the course of the treatment. Reported follow-ups ranged from 1 month to 21 years, 10.2% of the patients remained alive with the disease and 4.6% died secondary to the disease within two years of the diagnosis.

Recurrence of the tumor was encountered in 10% within a period ranging from 8 month to 13 years (Table 4).

4. Discussion

Endolymphatic sac tumor (ELST) is a rare neoplasm of the temporal bone. Although it was first recognized by Hassard et al. in 1984, the origin of this tumor has been debated until 1989 when Heffner et al. found that this neoplasm arose from the endolymphatic sac epithelium [5]. ELST has been underestimated and misinterpreted as paraganglioma, metastatic renal cell carcinoma, choroid plexus papilloma, ceruminous gland adenocarcinoma and aggressive papillary tumor of the temporal bone that was reclassified as ELST by Li et al. in 1993 [6].

The literature review showed an increase in the number of reported cases during the recent decade (Fig. 3). This could be attributed to the improvements in imaging and immunohistochemistry techniques (cytokeratine, vimentine, EMA and NSE), in addition to the fact that this tumor was recognized as a neuro-otologic manifestation of VHL disease in 1997 and screening for such an entity became mandatory in this group of patients [7].

VHL disease is an inherited, autosomal dominant phakomatosis with a variable expression (penetrance 95%) and prevalence of 1 per 39,000 people.

Patients with VHL disease have an increased risk of developing malignant or benign vascular tumors in various organs including central nervous system, pancreas, adrenals, kidneys and epididymis. This syndrome is related to mutations of the VHL suppressor gene located at the short arm of chromosome 3 (3p25–p26) that regulates transcription of vascular endothelial growth factor, the cell cycle, and extracellular matrix formation. There are more than 300 germline mutations in this gene that lead to overexpression of the vascular endothelial growth factor and subsequent development of vascular tumors [8,9].

The most common manifestations of VHL syndrome are retinal hemangioblastoma that appears first in the course of the disease, followed by cerebellar hemangioblastoma and renal cell carcinoma [10]. The incidence of ELST in VHL syndrome is around 15% in whom bilateral disease is encountered in one third of the cases [11]. Usually ELST occurs as a late manifestation of VHL syndrome but in our case (patient N 5) it was its first manifestation.

The patient had 15-month postoperatively new tumors appearance in the left cerebellum and cervical spinal cord that were resected and turned to be hemangioblastoma. Genetic studies confirmed the presence of VHL syndrome.

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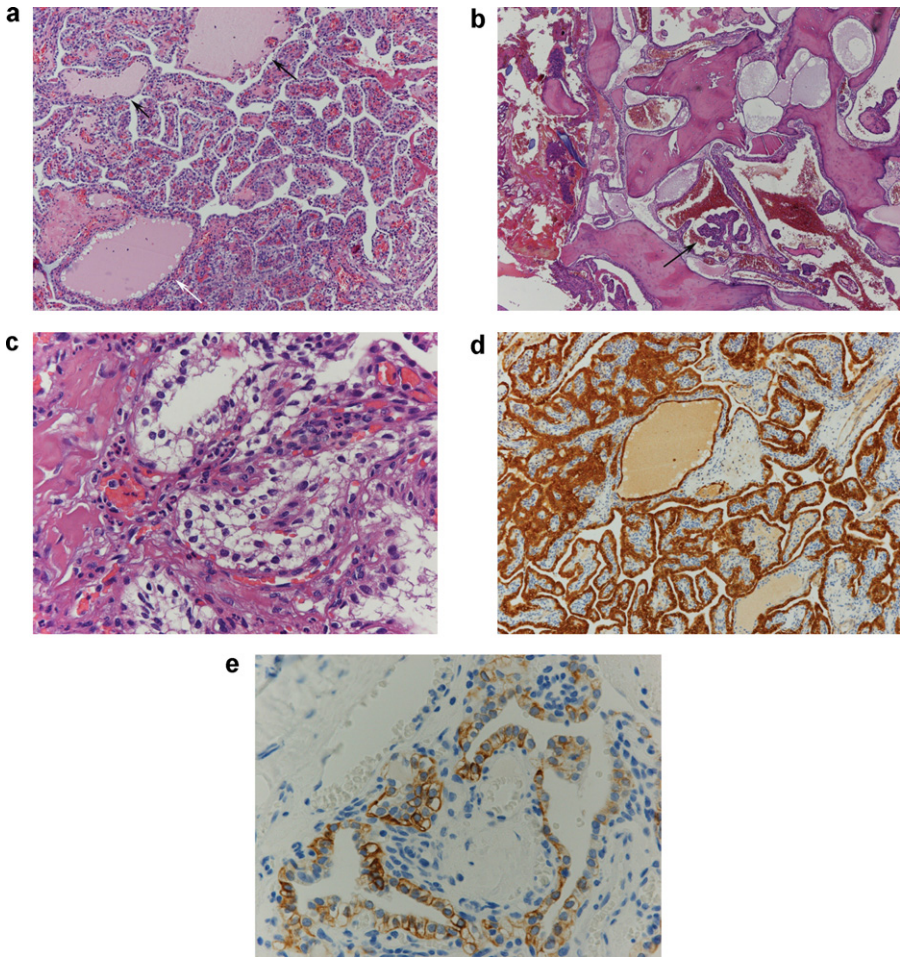

Fig. 2. (a) Prominent papillarity with a colloid-like cyst (white arrow) and two pseudocystic oedematous papillae (black arrows) (H&E, 100×). (b) Indolent bone infiltration with lamellae remodeling and focal papillary fronds (arrow) (H&E, 40×). (c) Clear cells with central or apical nuclei and capillaries just below the epithelium (H&E, 400×). (d) Intense immunoreactivity for NSE (100×). (e) Moderate immunoreactivity for GFAP (400×).

To the best of our knowledge, the occurrence of ELST as a late manifestation of VHL syndrome has been described only twice in the literature and only in children [10].

Hence, it is advisable to screen for VHL syndrome even in isolated cases of ELST. If genetic tests confirm the presence of VHL syndrome, the patients must have annually a brain and spine MRI, ophthalmoscopy, urinary catecholamine measurement, and an abdominal CT scan [8,9].

Prior to the diagnosis of ELST, six patients from our series complained of progressive hearing loss and the last patient presented with a sudden hearing loss of 2 months duration. The mechanism of hearing loss in ELST is explained by either a direct invasion of the inner ear or by an endolymphatic hydrops that mimic Meniere's disease. Endolymphatic hydrops can be confirmed by using the glycerol test and electrocochleogram [12].

Butman et al. explained the occurrence of sudden sensorineural hearing loss in ELST by an intralabyrinthine hemorrhage [13].

Diagnosing sporadic cases of ELST is challenging because of the nonspecific symptoms and difficult access for biopsy. Thus MRI and high resolution CT scan of the temporal bone are mandatory. In two large reviews Patel et al. and Mukherji et al. have found several radiological findings highly sensitive and specific for ELST:

1. Retrolabyrinthine location with local destruction centered at the posterior surface of the petrous bone.
2. Calcific speculations with posterior rim calcification (the site of origin) on CT scan.
3. Hyperintense focal signals on non enhanced T1-weighted MRI scan with heterogeneous enhancement post gadolinium.
4. Heterogeneous signal on T2-weighted MRI scan.

These findings are extremely useful to differentiate ELST from other destructive lesions of the temporal bone (vestibular schwannoma, chordoma, paragnaglioma and chondrosarcoma) [14].

On angiography, ELST appears as a moderately vascular tumor with irregular and not well defined tumor capsule. The reason of this heterogeneous vascularization pattern is the multiple small arterial supplies that derive from the

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Table 3
Summary of the demographic data and symptoms of ELST reported in the published series.

Author	P Nb	Age range and average	Sex	VHL	Side	T size (cm)	HL	Tin	FNP	ME mass	Vertigo	LCN deficit	Trig deficit	Interval time between tumor Dx and Sx (average)
Heffner (1989)	20	15–71 (41.4)	F: 10 M: 10	0	L: 11 R: 9	4–6	19	3	7	2	5	0	0	2–216 m (112 m)
Benecke (1990)	5	21–57 (39.4)	F: 4 M: 1	0	N/A	N/A	4	2	4	1	3	0	1	N/A
Feghali (1995)	3	19–46 (34)	F: 3 M: 0	0	L: 1 R: 2	N/A	3	2	2	2	0	1	0	8–12 m (10 m)
Megerian (1995)	8	36–71 (51)	F: 4 M: 4	1	L: 5 R: 3	N/A	8	6	3	5	4	2	1	6–276 m (128.2 m)
Roche (1998)	3	26–38 (30.6)	F: 2 M: 1	0	L: 2 R: 1	N/A	2	1	1	0	2	0	0	2–36 m (14.6 m)
Megerian (2002)	4	18–37 (27.5)	F: 3 M: 1	4	L: 2 R: 2	<2	4	4	0	0	2	0	0	N/A
Luff (2002)	3	24–68 47.3	F: 2 M: 1	0	L: 1 R: 2	2.7	3	2	1	0	1	0	0	120–180 m (160 m)
Rodrighes (2004)	7	29–66 (48.3)	F: 4 M: 3	0	L: 3 R: 4	N/A	6	3	5	3	3	1	1	N/A
Hansen (2004)	14	18–75 (42.4)	F: 10 M: 4	1	N/A	1–5.5	12	12	6	0	11	1	2	N/A
Schipper (2006)	7	36–58 (44.5)	F: 1 M: 6	1	N/A	N/A	6	N/A	0	N/A	N/A	N/A	N/A	N/A
Doherty (2007)	3	26–54 (35.3)	F: 2 M: 1	0	L: 2 R: 1	3.8–6.4	3	2	2	2	1	0	0	3–11 m (5.6 m)
Diaz (2007)	3	17–58 (31.6)	F: 2 M: 1	1	L: 3 R: 0	2–6	3	2	1	1	0	0	0	N/A
Ni (2008)	3	14–38 (31)	F: 1 M: 2	0	L: 1 R: 2	2.6–3	3	3	0	1	2	0	0	24–120 m (80 m)
Bae (2008)	4	15–67 (48)	F: 2 M: 2	0	L: 1 R: 3	3.2–4.5	4	0	2	0	2	0	0	N/A
Lonser (2008)	10	28–50 (39)	F: 4 M: 6	10	L: 9 R: 1	0.2–1.5	10	8	0	0	8	0	0	N/A
Cordreanu (2010)	3	16–46 (31.6)	F: 3 M: 0	3	L: 0 R: 3	N/A	2	0	0	2	1	0	0	8–72 m (40 m)
Our study	7	20–54 (30.4)	F: 5 M: 1	2	L: 2 R: 5	1.8–5	7	5	1	3	2	0	0	5–240 m (83 m)
Total	107	15–75 (38.1)	F: 62 M: 45	20.4%	L: 43 R: 38	0.2–6.4	94%	56%	32.7%	23%	47%	5%	5%	2–276 m (66.3 m)

Abbreviations: m, month; F, female; L, left; M, male; P, patient; R, right; T, tumor; Dx, diagnosis; FNP, facial nerve paralysis; LCN, lower cranial nerve; ME, middle ear; HL, hearing loss; N/A, not available; Nb, number,; Sx, symptoms; Tin, tinnitus; Trig, trigeminal nerve; VHL, von Hippel–Lindau disease.

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Table 4
Summary of recurrent cases of ELST reported in the published series.

Recurrent cases	Time from surgery	Preoperative RT	Treatment of recurrence	Status	Last F/U
Heffner et al.	12 m	+	Re-Operation	NED	48 m
Benecke et al.	108 m	–	Re-Operation (twice)	AWD	216 m
Feghali et al.	8 m	–	RT	AWD	48 m
Roche et al.	96 m	–	Re-Operation	NED	98 m
Hansen et al.	N/A	+	Re-Operation	AWD	261 m
Hansen et al.	N/A	–	Re-Operation	NED	61 m
Hansen et al.	12 m	+	Re-Operation (3 times)	DOD	39 m
Bae et al.	36 m	–	Re-Operation	NED	61 m
Cordreanu et al.	21 m	–	Re-Operation	AWD	29 m
Rodriguez et al.	N/A	+	Reoperated	NED	6 m
Our case	158 m	–	Re-Operation	NED	238 m

Abbreviations: m, month; AWD, alive with disease; DOD, dead of the disease; F/U, follow up; N/A, not available; NED, no evidence of disease; RT, radiotherapy.

ascending pharyngeal, middle meningeal, occipital, and anterior–inferior cerebellar arteries.

Five patients in our series with a tumor larger than 2 cm have undergone preoperative angiography with embolization. The devascularization effect was less than what is usually attained in temporal bone paragangliomas.

Based on imaging findings and tumor extension, Bambakidis et al. proposed an anatomical classification of ELST into four grades: grade I includes tumor limited only to the temporal bone, middle ear cavity, or external auditory canal, grade II includes tumor extending to the posterior fossa, and grade III includes tumor extending further into the middle cranial fossa. Once the tumor reaches the sphenoid wing or the clivus class IV is attained. In 2006 Schipper et al. suggested another but less detailed classification (Classes A–C) [15,16].

The mainstay treatment of ELST is surgical resection. All our patients underwent complete surgical removal of the tumor with preservation of the facial nerve.

The tumor abutted the facial nerve in three patients (patients 3, 4 and 6) and in one case (patient 5) the nerve was partially engulfed by the tumor with a preoperative grade 3 weakness.

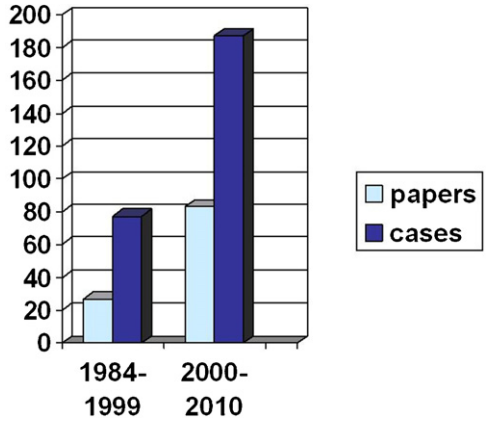

Fig. 3. This diagram shows an increase in the number of papers and reported cases of ELST during the past decade.

The management of the facial nerve was tailored according to the extension of the lesion and the degree of facial nerve involvement:

- Anterior rerouting was performed in 2 patients (cases 3 and 4) who developed mild to moderate facial weakness (grades 2 and 3)
- Posterior rerouting was done in one patient (case 5) in whom the facial nerve function did not change (grade 3).

Skeletonization of the nerve was achieved in one case (case 6) with normal postoperative facial nerve function.

Only two patients had worsening of their facial nerve function post operatively (Table 2).

In 3 patients the tumor had a mass effect on the middle cranial fossa dura. The area in contact with the tumor was coagulated with a bipolar for devitalization of any potential residual disease.

The involvement of the jugular bulb by the tumor was challenging due to its thin fragile wall and the proximity of the lower cranial nerves.

In 3 patients of our series the tumor infiltrated the jugular bulb. After confirming the patency of the contralateral cerebral vein system, the internal jugular vein was ligated in the neck first, the sigmoid sinus was packed (extraluminal and intraluminal) then the lateral wall of the jugular bulb dome was removed keeping intact the medial wall that protected the lower cranial nerves. Bleeding from the inferior petrosal sinus was controlled with Surgicell packing.

Butman et al. showed that hearing loss can occur even in very small tumor (2 mm) that spared the otic capsule where intralabyrinthine hemorrhage is responsible for an irreversible sudden sensorineural hearing loss [13]. Thus, in patients with evidence of even an asymptomatic ELST, a hearing salvage surgery, performed by an expert surgeon, should be taken into consideration. According to Megerian et al. the retrolabyrinthine transdural approach is the only hearing preservation surgery that ensure a total removal of the tumor with both the anterior and posterior sleeve of the

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dura, which could be potential sites for tumor recurrence [11].

The hearing was sacrificed in all our cases due to the advanced stage of the disease upon diagnosis. None of our patients has received adjunctive radiotherapy.

The analysis of the published series showed that 50% of the patients who received postoperative radiotherapy following subtotal resection of ELST had further growth of the residual tumor on follow up, and 20% of those who received radiation following complete removal of the disease had recurrence within one year. Consequently the role of postoperative radiotherapy (fractionated or stereotactic) in the treatment of ELST is still controversial.

The data showed also that patients with ELST had a less than 5% mortality rate and 10% recurrence rate. The patient number 6 had recurrence of the tumor 13 years following surgery, thus a long term follow up is mandatory after removal of the tumor. Seventy six percent of those patients who had residual tumor following subtotal resection or recurrence of the disease were still alive on last follow up (range from 10 m to 261 months).

No reported case of distant haematogenous metastasis was encountered in the literature. Drop metastasis was reported in two patients, one to the spinal cord and the other to the cerebellum [3].

5. Conclusion

Complete surgical resection of ELST is the treatment of choice and preoperative embolization is beneficial in large tumors. Hearing preservation surgery could be successful only in the early stage of the disease and should be attempted specially in VHL syndrome in which bilateral tumor is found in 5% of the cases.

Although ELST is histologically a low grade malignant tumor, it has relatively a good prognosis. Thus the facial nerve should not be sacrificed unless it is totally invaded by the tumor. Postoperative adjunctive radiotherapy plays a limited role in the treatment of ELST and long term follow up is mandatory.

Conflict of interest

None of the authors has any conflicts of interest to disclose nor did the study receive any external funding.

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