

Case Report

Gamma knife radiosurgery in the management of endolymphatic sac tumors

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Abstract

Background: Although widely regarded as rare epithelial tumors with a low grade of malignancy, endolymphatic sac tumors (ELST) often lead to disabling petrous bone destruction and significantly impairing symptoms at the time of primary diagnosis and/or recurrence. ELST is not uncommon in von Hippel Lindau (VHL) patients. Although open surgery is regarded as the best treatment option, recurrence remains a challenge, particularly when gross tumor resection (GTR) is deemed unachievable due to topographic conditions. Tumor recurrence successfully treated with fractionated radiotherapy and radiosurgery have been reported in selected cases. We present the case of a patient with recurrent ELST treated with salvage gamma knife radiosurgery (GKRS) adding a review of current literature.

Case Description: A 65-year-old patient underwent GKRS of an unresectable, recurrent ELST. Tumor volumetric analysis showed almost 15% increase in tumor volume in the 4 months between the pre-GKRS magnetic resonance imaging (MRI) and the stereotactic MRI (s-MRI) at treatment. Follow-up MRI at 12 and 20 months showed significant decrease in local tumor volume, decreased contrast enhancement and no perifocal edema. The patient's general and neurological status remains stable to the present day.

Conclusion: In the present case, GKRS was effective in the management of a recurrent ELST over the course of 20 months. Because of ELSTs recurrence potential, long-term follow up is required. The present case as well as previous reports might suggest a possible salvage/adjunctive role of radiosurgery in the management of ELST. Further studies are deemed necessary.

Key Words: Endolymphatic sac tumors, gamma knife radiosurgery, tumor invasiveness, von Hippel Lindau

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INTRODUCTION

Endolymphatic sac tumors (ELST) are rare neoplasms derived from the endolymphatic sac of the inner ear. They are slow growing tumors with a low grade of malignancy, but are locally aggressive and destructive.^[21] Although total surgical resection is the treatment of choice, it is not always feasible which can lead to post-operative recurrence.^[9,11,26] Fractionated radiotherapy and stereotactic radiosurgery have been reported as treatment options in a limited number of cases.

This case report describes a sporadic ELST that recurred after three surgical resections and was treated by gamma knife radiosurgery.

CASE PRESENTATION

We present the case of a previously healthy, now 67-year-old female patient developing vertigo, nystagmus, wide-based gait, and memory loss throughout July and August 1999 (49 years of age at that time). A Computed Tomography (CT) scan of the brain (August 1999) revealed a 35-mm macrolobulated, extraaxial, brightly contrast enhancing mass in the left posterior fossa with extension along the petrous temporal bone, tentorium cerebelli and sigmoid sinus, with cerebellar edema, compression of the fourth ventricle, supratentorial hydrocephalus as well as erosion of the petrous temporal bone. The tumor was radiologically assessed as a meningioma. Subtotal resection (STR) and ventricular drain placement were performed shortly thereafter (Sept 1999). Audiograms prior and after this surgical intervention are currently not available for review. The patient's condition improved after surgery (pre-operative KPS = 70, post-operative KPS = 90). The initial microscopic evaluation proved complex; after major scrutiny, the pathological report described the tumor as a fibroblastic meningioma.

The patient was then lost to follow-up until readmitted six years later (2005) for unilateral progressive hearing loss and pressure like symptoms in her left ear. A clinical examination revealed abnormal bulging of the left tympanic membrane and conductive hearing loss assessed as Gardner-Robertson grade I. A new CT scan and a corresponding MRI (May 2005) showed heterogeneous contrast enhancement suspicious of a local recurrence. The patient underwent a second surgery (August 2005) again assessed as STR. The histopathological examination revealed this time an endolymphatic sac tumor (ELST). Available medical data showed no evidence of von Hippel Lindau (VHL) disease screening prior to both surgeries; to our knowledge, the patient has no known criteria for VHL disease.

Follow-up imaging between 2006 and 2008 showed gradual, but very slow increase in size of a suspected

Table 1: Microscopic profile (MP) at first and third surgery. MP at second surgery (2005) not available

Microscopic evaluation	At first surgery (1999)	At third surgery (2009)
Cytokeratin MNF 116	Positive	Positive
CD 34	Positive	Positive
EMA	Positive	Positive
Progesterone receptor	Positive	Positive
Chromogranin	Negative	Negative
Synaptophysin	Negative	Negative
TTF-1	Negative	Negative
Stat6	Negative	Negative
P63	Negative	Negative
Calponin	Negative	Negative

Intrinsic proliferative activity (Ki67) not included in the above analysis as the majority of Ki67 immunoreactive nuclei corresponded to tumor-infiltrating inflammatory cells

Table 2: Audiogram evolution throughout all surgical interventions (1999-2017)

	GB-scale	PTA	SD
Audiogram at post surgery nr 1 (1999)	NA	NA	NA
Audiogram at post-surgery nr 2 (May 2005)	I	23 db	NA
Audiogram post-surgery 3 (October 2009)	II	44 db	96%
Audiogram at post-GKRS (July 2017)	III	53 db	NA

GB: Gardner Robertson, PTA: Pure Tone Audiometry, SD: Speech discrimination

Table 3: Tumor volume values prior (pre-GKRS), at treatment and after GKRS (post-GKRS)

Date	Tumor volume (cm ³)	Tumor size evolution (%)
Pre GKRS MRI (October 2014)	4.43	-
stereotactic-MRI (February 2015)	5.15	↑16%
Post GKRS (February 2016, 12m)	3.87	↓25%
Post GKRS (November 2016, 18m)	1.69	↓56%

residual/recurrent tumor. A follow-up MRI in February 2009 confirmed a local recurrence in the left sigmoid sinus region and the patient underwent a third surgery (STR) in February 2009. The corresponding pre-operative audiogram is currently not available. The histopathology confirmed an ELST [Table 1; Figure 1a, b and c]. The microscopic re-evaluation of all samples collected at first surgery (1999) proved consistent with ELST [Table 1]. A post-operative audiogram in August 2009 showed further hearing deterioration (Garden-Robertson II) [Table 2].

MRI examinations from April 2009 to August 2011 demonstrated no evidence of focal recurrence. Follow-up MRI scans from 2012 to 2013 are currently not available for review. Follow-up MRI in October 2014 showed a local, aggressive recurrence within the lateral limits of the surgical region [Figure 2a]; clinical evaluation demonstrated further (partial) hearing loss on the left side without any further cranial nerve dysfunction;

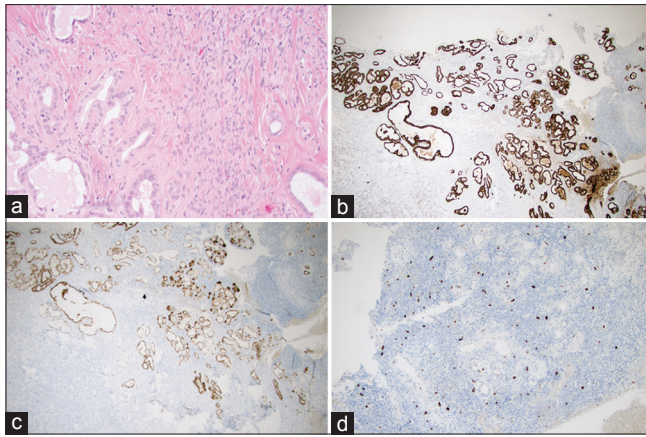


Figure 1: (a–d) The microscopic architecture of ELST at third surgery (Magnification 200×). (a) Hematoxylin and eosin (H and E) staining; (b) CK-MNF immunohistochemical staining positive in glands; (c) EMA immunohistochemical staining positive in glands; (d) Ki67 immunohistochemical staining for proliferative cells

unfortunately, the corresponding audiogram is currently not available. A fourth microsurgical intervention was assessed not indicated at this point. Gamma Knife radiosurgery (GKRS) was assessed as the best surgical option. The patient underwent single session GKRS-treatment (Perfexion™ Gamma Knife model) in February 2015 (72 months after the third and last microsurgery). Fixation was achieved using the Leksell® Coordinate Frame G (Elekta AB, Stockholm). Leksell GammaPlan® (LGP)-gross tumor volume (GTV) assessment proved a substantial increase in tumor volume of approximately 15% between the pre-GKRS MRI (4.43 cc - October 2014) and the stereotactic-MRI (s-MRI) at treatment (5.15cc - February 2015). The tumor was best delineated by the fat-saturated T1 contrast-enhanced s-MRI sequence; the peripheral dose was set at 18Gy on the 50% isodose line. GTV margins were equal to clinical target volume (CTV) [Figure 2b]; planning target volume (PTV) margins were not required. The overall procedure was well tolerated by the patient.

RESULTS

Post-GKRS MRI at 12 months showed a decrease in tumor volume of 25% (3.87cm³) with no evidence of adverse radiation event. A further follow-up MRI at 20 months (November 2016) showed a further decrease of the lesion's volume [Figure 2c-d], measuring 1.69 cm³ (67% tumor volume reduction compared to s-MRI February 2015) [Table 3]. Post-GKRS audiogram performed in July 2017 showed further hearing deterioration assessed as Gardner-Robertson grade III [Table 2]. The patient's general and neurological condition remained otherwise stable at the time of paper submission (KPS 100, no other neurological impairment).

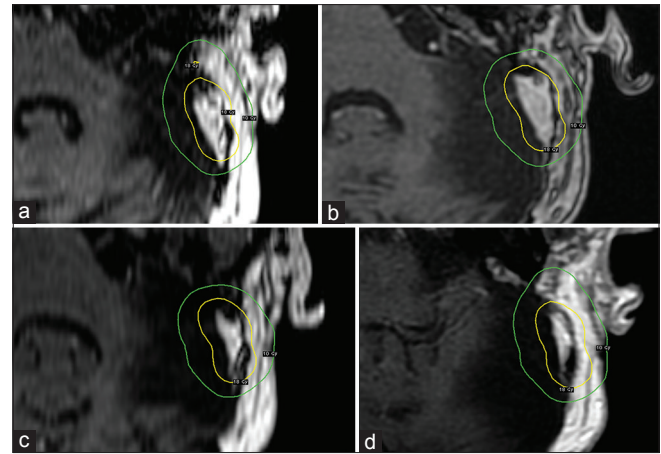


Figure 2: (a–d) MRI of the tumor; (a) Pre GKRS MRI, October 2014. Tumor volume 4.43 cm³. (b) Stereotactic (Treatment) MRI, Feb. 2015. Tumor volume 5.15 cm³. (c) Post GKRS MRI at 12 months, Feb 2016. Tumor volume 3.87 cm³. (d) Post GKRS MRI at 20 months, Nov 2016. Tumor volume 1.69 cm³

DISCUSSION

ELSTs might present with a complex evolution due to their aggressive erosive growth pattern and recurrence potential. The diagnostics of ELSTs require multidisciplinary assessment; yet, misdiagnoses may still take place as illustrated in our case. We will discuss the traits common to ELSTs in terms of clinical features, diagnostics and available treatments.

Clinical features

ELST was first reported in 1984 after a sac decompression of a presumed unilateral Ménière's disease.^[6,12] Due to their localization and aggressive erosive character, patients may develop serious impairing symptoms by the time of diagnosis or recurrence. Although not the case here, ELSTs are found in 10% of the VHL population.^[2,9] As in our case, most of ELSTs are isolated/sporadic (unilateral) tumors, whereas bilateral cases are mostly seen in patients with VHL syndrome. Sporadic ELST seems to have a more aggressive evolution,^[17,23] which correlates well to the present case. As in our patient, audiovestibular impairment is often present by the time of diagnosis.^[4,9] Patients commonly develop unilateral sensorineural hearing loss; ipsilateral facial nerve paresis, otalgia, otorrhea, tinnitus, vertigo, headache, and ataxia are also common features.^[5,9] As in our case, hydrocephalus may also develop. Depending on tumor extension, dysfunction of other cranial nerves (including Vernet's syndrome and trigeminal symptoms) may also arise.^[5,7] Distant dissemination is very uncommon but a few metastatic cases have been reported.^[2,3,5,9,22]

Microscopic features

According to the WHO, ELSTs are defined as low-grade malignant epithelial tumors of endolymphatic sac origin.^[21] Because of ELSTs' complex microscopic

features, the differential diagnosis includes schwannoma, meningioma (including malignant meningioma with adenocarcinoma-like metaplasia), jugular paraganglioma, choroid plexus papillomas, solitary fibrous tumor with salivary gland inclusion as well as metastases from papillary thyroid and renal cell carcinomas.^[5,6,9] In our case, meningioma proved to be the main differential concern.

ELSTs are microscopically composed of papillary fronds and thyroid follicle-like glandular structures. Hemorrhage, calcifications, siderophages, cholesterol clefts, and inflammatory cells are frequently described.^[5] The immunohistochemical profile is usually positive for cytokeratin (types 5,6,7,8,17,19), Epithelial Membrane Antigen (EMA), Vimentin, and Periodic acid-schiff; ELSTs may also stain positive for Neuron specific enolase (NSE), S100, and Glial Fibrillar Acid Protein (GFAP).^[6,9] Transthyretin can be effective to differentiate ELSTs from choroid plexus papillomas; its absence or very limited expression in collected samples points to ELSTs.^[6] Metastatic lesions from papillary thyroid cancer could be identified by positive thyroglobulin and TTF1 immunohistochemistry reaction.^[6] Malignant morphological features in both glands and stroma could indicate the evolution of malignant meningioma with adenocarcinoma like metaplasia; yet, as in our case, ELST usually displays uniform glands with uniform epithelial features.^[18] As previously mentioned, other differential diagnoses include solitary fibrous tumor with salivary gland inclusion; in this context, STAT6 (negative in ELSTs) is a reliable histochemical marker and the features of cells are follicles rather than acinar or serous glands.^[20] Although rare tumors, myoepitheliomas of the bone may present misleading characteristics; these tumors show myoepithelial differentiation rather than ductal differentiation. They are positive for cytokeratin and EMA in conjunction with S100 protein and myogenic markers such as SMA and calponin; other markers include GFAP and p63. EWSR1 gene rearrangement may also be found.^[13] In our case, Calponin and p63 proved negative. Myoepitheliomas of the middle ear could also be included in the much rarer spectrum of differential diagnoses.^[13] Table 2 describes the available microscopic evaluation (MP) at each surgery for our case.

Neuroimaging

State-of-the-art neuroimaging, including vascular imaging is critical for the diagnosis and management of ELSTs, including MRI, CT with thin slice bone algorithm and digital subtraction angiography (DSA). High-resolution CT often demonstrates a lytic bone lesion in the endolymphatic sac region, effectively delineating the extent of bony erosion which usually involves the retrocochlear posteromedial border of the petrous temporal bone; central spiculated calcifications are also commonly described.^[5] On the MRI, multiple

high-signal intensity foci on unenhanced both T1- and T2-weighted images indicate the presence of blood, methemoglobin, protein-filled cysts or cholesterol clefts, which are uncommon features in other temporal bone tumors,^[9,10] with a differential diagnosis including the typically more homogeneously high T1 and T2 signal of cholesterol granuloma, a non-neoplastic lesion located classically more medially in the petrous apex. T1-weighted gadolinium-enhanced sequences often show heterogeneous enhancement mainly due to the tumor's level of vascularity.^[5] DSA is performed to display the tumor's vascular structure with its often complicated blood supply;^[5] it is used to map pre-surgical landmarks as well as to differentiate ELST from other differential diagnoses. ELSTs may display multiple/complex blood supply from the external and internal carotid arteries as well as the vertebrobasilar system; sole supply by the internal carotid or posterior circulation is uncommon due to ELSTs anatomical origins.^[6]

Treatment

Early GTR is widely regarded as the treatment of choice as it often leads to long disease-free intervals.^[6,9,11,16,17,26] A large number of surgical procedures are available according to the tumor size and extension^[5,19]. Nevertheless, GTR is often unachievable due to local anatomical factors^[9,10] such as cranial nerve involvement and focal complex vascular supply and drainage. As such, successful resections are commonly dependent on the use of advanced methods such as preoperative angiography/embolization and intraoperative electrophysiological monitoring.^[5] The role of adjuvant radiotherapy, including GKRS, has been restricted by the limited available data.^[5,25] Rodney *et al.* (2007) described two patients with ELST who underwent GTR whilst a third case after STR retained microscopic residual tumor adjacent to the neural structures of the medial jugular foramen and the internal carotid artery.^[6] In this series, all ELST patients received adjuvant radiotherapy (two patients with fractionated external beam radiotherapy and one with proton beam therapy). Post-surgical MRI surveillance (follow-up intervals of 10–144 months) showed progression-free conditions in all patients. The group also reported sporadic use of adjunctive stereotactic radiosurgery after STR, but each of these had led to tumor recurrence.^[6] Carlson *et al.* reported that STR carried a significant risk of recurrence, thus the authors strongly recommended early GTR.^[4] The same group also described the use of SRS on an unresected tumor achieving local tumor control at last follow-up (94 months). The authors concluded that SRS should be considered when primary or salvage surgery are assessed as not feasible or contraindicated.^[9] Yet, Nevoux *et al.*^[17] reported four cases of local recurrence after GTR on a series of 14 ELST patients (28%) under a follow-up period of 14 years, underscoring the importance of long-term follow-up and the value of adjunctive treatment radiotherapy. Kunzel *et al.* described a case of ELST successfully

treated with surgery and post-operative fractionated radiotherapy (up to 60 Gy), with a 10-year follow up.^[16] Yu *et al.* (2011) reported similar results on a patient undergoing complementary fractionated radiotherapy (LINAC) of up to 50.4 Gy after STR; no signs of recurrence or side effects/toxicity were observed after one year follow up; long-term follow up was also strongly suggested.^[27] Despite the above, other groups have questioned the efficacy of post-operative conventional fractionated radiotherapy due to the significant number of recurrences reported at post surgery (up to 50% after STR and 20% after GTR); the same groups suggested instead the use of radiosurgery in poor surgical candidates or when the morbidity of salvage surgery is deemed significant.^[19] A number of publications seem to support the latter. Cheng *et al.*^[5] described a case of bilateral multifocal, recurrent ELSTs treated with four different GKRS sessions; the authors described long term control of all tumors after delivering peripheral doses of 11 to 16 Gy. Balasubramaniam *et al.*^[11] described a case of ELST-recurrence in the region of the right jugular foramen 2-years post-resection; GKRS (15 Gy) achieved positive clinical and radiological evolution 2.5 years post GKRS. Kamida^[14] described similar results in a case with ELST delivering 21 Gy. Virk *et al.* (2013) also described the use of GKRS in a case of ELST recurrence; however, in this case, the patient had to undergo subtotal petrosectomy due to post GKRS local failure at a later stage. The prescription dose and the time interval between GKRS and the petrosectomy were not specified.^[25] According to our review of literature, GKRS seems to provide tumor control in selected ELST cases. The latter observation seems congruent with the reported effects of radiosurgery in other low-grade malignant tumors of the head and neck, particularly salivary gland carcinomas.^[7,8,15,24]

CONCLUSION

Despite being low-grade malignant tumors, ELSTs may present a complex clinical and radiological evolution. Early GTR is widely regarded as first hand treatment but is not always feasible due to local topographic conditions. Cases of recurrence after surgery have been reported and are mainly associated with STR; however, recurrence after GTR may also occur. Repeated resection at the time of recurrence might also prove hazardous due to the previously discussed factors. In these cases, salvage GKRS might have a positive impact in terms of tumor control/tumor progression-free survival; seemingly, good long term results may be achieved prescribing a peripheral dose of 15–18 Gy. Nonetheless, regardless of the treatment modalities chosen and evident positive outcome, we recommend long-term follow up in this group of patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information

to be reported in the journal. The patient understands that name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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