

# Endolymphatic sac tumor – case report

## Guz woreczka śródchłonki – opis przypadku

### Authors' Contribution:

A – Study Design  
B – Data Collection  
C – Statistical Analysis  
D – Manuscript Preparation  
E – Literature Search  
F – Funds Collection

Jerzy Kuczkowski<sup>1ABDE</sup> Wojciech Brzoznowski<sup>1BEF</sup> Tomasz Nowicki<sup>2B</sup> Jolanta Szade<sup>3B</sup>

<sup>1</sup>Klinika Otolaryngologii Gdański Uniwersytet Medyczny, Kierownik: prof. dr hab. Czesław Stankiewicz<sup>2</sup>

<sup>2</sup>II Zakład Radiologii Gdański Uniwersytet Medyczny, Kierownik: dr hab. med. Edyta Szurowska prof. Gdański Uniwersytet Medyczny

<sup>3</sup>Zakład Patomorfologii Gdański Uniwersytet Medyczny, Kierownik: prof. dr hab. Wojciech Biernat

Article history: Received: 17.01.2017 Accepted: 30.01.2017 Published: 30.03.2017

### ABSTRACT:

The aim of this paper is to present the case of a 70-year-old woman with endolymphatic sac tumor and temporal bone destruction treated at Otolaryngology Department of MUG. The patient was admitted to our Department due to a 3-year history of hearing loss, dizziness and ear pain. The first diagnosis was temporal bone tumor connected with von Hippel-Lindau syndrome (VHL). The patient was surgically treated. During intraoperative examination, a neoplasm was determined. The tumor was excised via transmastoid approach with sigmoid sinus skeletonization. After treatment, her pains disappeared. Histopathological and immunohistochemical examination revealed endolymphatic sac tumor. Follow-up CT showed no tumor remission.

### KEYWORDS:

temporal bone, tumors, ELST, management

### STRESZCZENIE:

Celem pracy jest omówienie przypadku guza woreczka endolimfatycznego z destrukcją ucha wewnętrznego u 70-letniej kobiety leczonej w Klinice Otolaryngologii GUMed. Chora została przyjęta do Kliniki z powodu bólów ucha, głuchoty, zawrotów głowy trwających od trzech lat. Postawiono wstępne rozpoznanie guza rozwijającego się w kości skroniowej w przebiegu zespołu von Hippel-Lindau (VHL). Chorą leczono chirurgicznie. W badaniu doraźnym stwierdzono obecność nowotworu. Wykonano operację usunięcia guza z dojścia przesuskowego ze szkieletowaniem zatoki esowatej. Po zabiegu dolegliwości bólowe ustąpiły. Badania histopatologiczne oraz immunohistochemiczne wykazały obecność guza woreczka śródchłonki (ELST). Kontrolne badanie TK nie wykazało wznowy guza.

**SŁOWA KLUCZOWE:** kość skroniowa, nowotwory, ELST, postępowanie

## INTRODUCTION

Endolymphatic sac tumor (ELST) is a very rare neoplasm of the inner ear developing from neuroectoderm. The first description of ELST comes from 1984 when a small tumor was accidentally found during decompression of the endolymphatic sac (due to Ménière's disease). [1] In the literature, those tumors were earlier classified as aggressive papillary middle ear tumors (APMET). However, it was not until 1989 when Heffner presented ELST as a separate disease – a tumor developing from epithelium of the endolymphatic sac, what he called a low-grade adenocarcinoma. [2] Since then, it has often been referred to as Heffner's tumor. In 1993, Li et al. reclassified

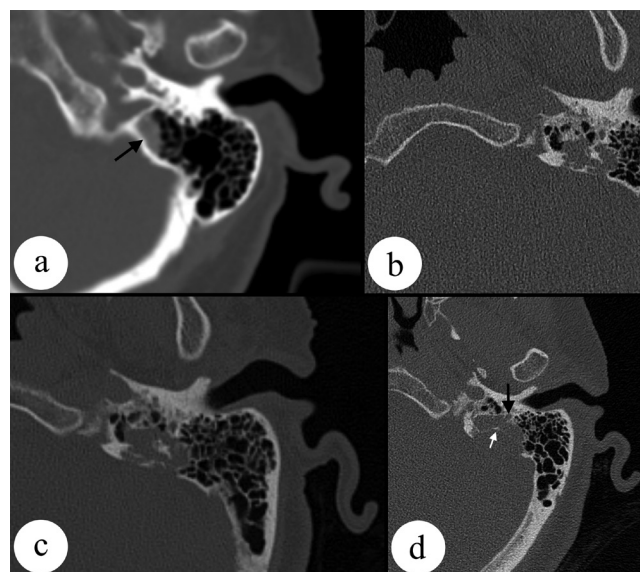
those tumors, naming them as ELST. [3] Discovery of immunohistochemical markers significantly contributed to classification of this group of tumors. [4] Approximately 200 cases of this tumor have been reported in the literature thus far. Most of them occur sporadically, however, some of them are related to a rare genetic condition called von Hippel-Lindau disease (VHL) caused by mutation of chromosome 3 (3p25-26). [5-6] The prevalence of ELST in patients suffering from VHL syndrome is ca. 10%, while in general adult population it amounts to 1: 30000. [7] Although it is a histologically benign tumor, it can be locally aggressive leading to destruction of surrounding structures. Symptoms of ELST include ear pain, progressing hearing loss and vertigo. Diagnosis is based on

audiometry, ENG, head CT and MRI as well as pathology study. Differential diagnosis should include chromaffin-negative paraganglioma, cholesterol granuloma and chronic granulomatous otitis media or metastatic adenocarcinoma. Surgical treatment depends on clinical stage. We present a case of the patient with ELST in the course of von Hippel-Lindau disease treated in our Department.

## CASE REPORT

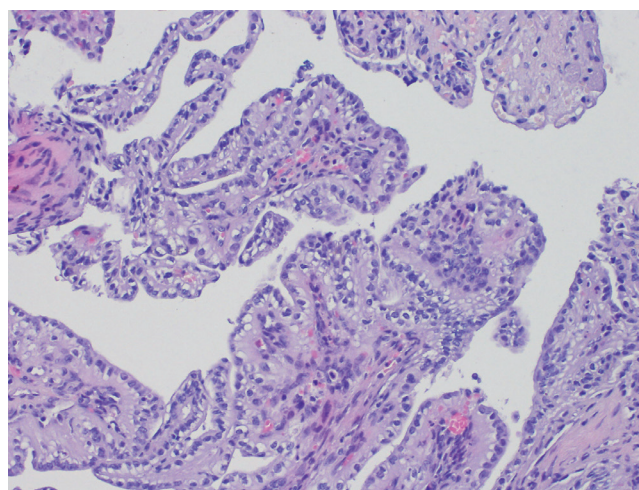
A 70-year-old female patient was admitted to the Department due to a tumor of the left temporal bone. For three years, she had been treated for progressing headaches of the left temporal and parietal regions, vertigo with imbalance. Additionally, the patient complained of tinnitus with rapidly progressing hearing loss on the left side for one year. The function of the facial nerve was preserved. In the history, numerous surgical interventions due to neoplastic diseases were noticeable (thyroidectomy for multinodular goiter, gastrectomy for stromal GIST tumor, left-sided mastectomy for breast cancer, hysterectomy for endometrial cancer). Other comorbidities included: acromegaly, hypothyroidism, paroxysmal atrial fibrillation, symptomatic bradycardia (requiring electrostimulation), hypertension. On the otoscopy examination, the tympanic membrane was pearl-grey and unaffected on both sides. Strong soreness of the planum mastoideum and apex of the mastoid process of the left ear was noted. Audiometry revealed hearing loss on the left side. No nystagmus. On VNG examination – trace of excitability. On a CT scan of the temporal bone (prior to surgery) – destruction of the posterior wall of the petrous part of the left temporal bone and destruction of the vertical part of the facial canal, internal auditory meatus, posterior semicircular canal, crus commune of the semicircular canals. Within the lesion, spiculated calcifications were visualized.

Considering general condition and anesthesiological limitations, the patient underwent extended mastoidectomy. Intraoperatively, a cystic tumor was found that was grey-blue in color and located in the apex of the mastoid process, meninges of the posterior cranial fossa, sigmoid sinus, infiltrating towards the labyrinth. The tumor was resected from the apex of the mastoid process, sigmoid sinus wall and posterior petrous part of the temporal bone (Moritz-Trautmann triangle). During the procedure, no excessive bleeding or cerebrospinal fluid leakage were noted. The postoperative course was uncomplicated. Slight vertigo pertained with tendency to fade spontaneously. After the intervention, the headaches resolved completely. Pathology study revealed endolymphatic sac tumor with the following phenotype: EMA+, CK AE1/3+, GFAP -/+, thyroglobulin -, TTF -.



**Fig. 1.** CT temporal bone in a patient with ELST in different periods of the disease (before surgery).

- a) High resolution CT of left temporal bone in axial plane. The lesion with destruction of the posterior aspect of the temporal bone is visible.
- b) ..... High resolution CT of left temporal bone in axial plane. The lesion with destruction of the posterior aspect of the temporal bone is visible.
- c) Follow-up CT examination in bone window one year after the previous examination. Further enlargement of the lesion can be seen.
- d) High resolution CT of left temporal bone performed before operation reveals a massive destruction of bone structures with complete loss of posterior aspect of temporal pyramid and partial destruction of walls of vertical part of facial canal. Within the lesion a typical speculated calcification is visible.



**Fig. 2.** Histopathological investigation of the endolymphatic sac tumor. The papillary structures were lined by a single layer of flattened cuboidal-to-columnar cells with clear cytoplasm and minimal pleomorphism. (H&E, 200×).

After oncologic consultation, it was decided not to introduce adjuvant radiotherapy. On control CT scan of temporal bones (6 months after operation), the tympanic cavity was normally aired, postoperative cavity in the mastoid part filled with connective tissue scar.

## DISCUSSION

The most common symptom of the endolymphatic sac tumor is vestibulocochlear dysfunction. Almost all patients present asymmetric sensorineural hearing loss of varying degree, which corresponds with the extent of the tumor. [8] Progressing sensorineural hearing loss accompanied by tinnitus observed in this case could have been caused by neoplastic infiltration to the bony labyrinth or as a result of developing endolymphatic hydrops. [9] Imbalance and sensation of fullness in the ear are other typical symptoms of this tumor (70% and 37% of patients respectively). [9] Growing tumor causes obstruction of endolymphatic reabsorption and excessive production of fluid, which both contribute to development of secondary endolymphatic hydrops. The previously mentioned symptoms can mimic Ménière's disease, which postpones correct diagnosis. It illustrates the need to diagnose Ménière's disease by exclusion, where diagnostic imaging (CT/MRI) should be compulsory in order to exclude other pathologies. Endolymphatic sac tumor is initially localized on the posteromedial surface of the petrous part and it later grows towards cerebellopontine angle and posterior cranial fossa. It is important to differentiate it from chromaffin-negative paraganglioma, meningioma, schwannoma, cholesterol granuloma, middle ear adenoma or metastases of thyroid cancer, kidney, prostate, lung or breast cancer. After reaching sufficient size, the tumor presses the brainstem, which exacerbates vestibular symptoms and headaches. Second most common pathway of neoplastic growth leads laterally through middle ear and mastoid process, usually involving facial nerve or generating symptoms resembling

chronic otitis media with Eustachian tube dysfunction. [10] For diagnosis and treatment planning of ELST, CT and MRI scans are useful. Those studies reveal a soft tissue tumor with characteristic spiculated calcifications and a thin ring along the posterior surface of the tumor. On the CT scan performed in an out-patient setting in this patient 2 years ago, a lesion with well-defined borders and density of soft tissue was found in the medial portion of the mastoid process (Fig.1a). This lesion was not included in the description and therefore the patient was not further managed. In the case of such tumor, it is indicated to perform angiography. Those tumors are richly vascularized and preoperative embolization of the main artery (usually it is the ascending pharyngeal artery or posterior auricular artery) limits intraoperative bleeding. The best results are obtained in the case of well vascularized tumors greater than 2cm in size. In our case, it was not necessary to perform this study. The treatment of choice of ELST is surgery. The tumor is resected locally during mastoidectomy or petrosectomy. In some cases, adjuvant radiotherapy is indicated. The greatest chances of radical removal are achieved at the early stage of the disease. Extensive lesions are sometimes inoperable due to involvement of crucial structures (e.g. brainstem). In the case of dura mater involvement, it should be excised and the defect should be filled with musculofascial flap of the temporal muscle. Temporal bone defect should be obliterated with fat. Large hopes are related to Gamma Knife treatment. [11]

## CONCLUSIONS

- Endolymphatic sac tumors are rare tumors of the temporal bone, which can cause severe headaches and lead to hearing loss.
- Diagnosis is based on diagnostic imaging (CT/MRI), audiometry and pathology study.
- Treatment of choice of endolymphatic sac tumors is surgical resection.

## References

1. Hassard A.D., Boudreau S.F., Cron C.C.: Adenoma of the endolymphatic sac. *J. Otolaryngol.* 1984; 13: 213–216.
2. Heffner D.K.: Low-grade adenocarcinoma of probable endolymphatic sac origin: a clinicopathologic study of 20 cases. *Cancer.* 1989; 64: 2292–2302.
3. Li J.C., Brackmann D.E., Lo W.W., Carberry J.N., House J.W.: The reclassification of aggressive adenomatous mastoid neoplasms endolymphatic sac tumors. *Laryngoscope.* 1993; 103: 1342–1348.
4. Turner J., Chang P., Noushi F., Atlas M., Fagan P.: Aggressive papillary tumor of the temporal bone: new immunohistochemical evidence for endolymphatic sac origin. *Austral. J. Otolaryngol.* 1998; 3:50–58.
5. Megerian C.A., McKenna M.J., Nuss R.C., Maniglia A.J., Ojemann R.G., Pilch B.Z., Nadol J.B. Jr.: Endolymphatic sac tumors: histopathologic confirmation, clinical characterization, and implication in von Hippel-Lindau disease. *Laryngoscope.* 1995; 105: 801–808.
6. Maher E. R., Kaelin W.G. Jr.: Von Hippel-Lindau disease. *Medicine.* 1997; 76: 381–391.

7. Manski T.J., Heffner D.K., Glenn G.M., Patronas N.J., Pikus A.T., Katz D. et al.: Endolymphatic sac tumors: a source of morbid hearing loss in von Hippel-Lindau disease. *J. Am. Med. Assoc.* 1997; 277: 1461–1466.
8. Bambakidis N.C., Megerian C.A., Ratcheson R.A.: Differential grading of endolymphatic sac tumor extension by virtue of von Hippel-Lindau disease status. *Otol. Neurotol.* 2004; 25: 773–781.
9. Butman J.A., Kim H.J., Baggenstos M., Ammerman J.M., Dambrosia J., Patsalides A. et al.: Mechanism of morbid hearing loss associated with tumors of the endolymphatic sac in von Hippel-Lindau disease. *JAMA.* 2007; 298 (1): 41–48.
10. Mishra G., Sharma Y., Padhya C., Parikh B., Gupta M. et al.: Endolymphatic duct papillary tumor: capture dundercover of complicated chronic otitis media. *Indian J. Otolaryngol. Head Neck Surg.* 2013; 65: 662–664.
11. Ferreira M.A., Feiz-Erfan I., Zabramski J.M., Spetzler R.F., Coons S.W., Preul M.C. et al.: Endolymphatic sac tumor: unique features of two cases and review of the literature. *Acta Neurochir. (Wien).* 2002; 144: 1047–1053.

---

Word count: 1400   Tables: —   Figures: 2   References: 11

---

Access the article online:   DOI: 10.5604/01.3001.0009.7986

---

Table of content: <http://otorhinolaryngologypl.com/resources/html/articlesList?issuelid=9790>

---

Corresponding author: Jerzy Kuczkowski; Klinika Otolaryngologii Gdański Uniwersytet Medyczny, Polska; e-mail: [jerzyk@gumed.edu.pl](mailto:jerzyk@gumed.edu.pl)

---

Copyright © 2017 Polish Society of Otorhinolaryngologists Head and Neck Surgeons. Published by Index Copernicus Sp. z o.o. All rights reserved

---

Competing interests: The authors declare that they have no competing interests.

---

Cite this article as: Kuczkowski J., Brzoznowski W., Nowicki T., Szade J.: Endolymphatic sac tumor – case report; *Pol Otorhino Rev* 2017; 6(1): 39-42

---