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# Endolymphatic Sac Tumor Showing Increased Activity on $^{68}\text{Ga}$ DOTATATE PET/CT

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**Abstract:** Endolymphatic sac tumors (ELSTs) are rare tumors arising from the epithelium of the endolymphatic sac and duct that can be either sporadic or associated with von Hippel-Lindau (VHL) disease. We report a case of a VHL patient with histologically proven residual ELST who underwent  $^{68}\text{Ga}$  DOTATATE PET/CT showing increased activity ( $\text{SUV}_{\text{max}}$ , 6.29) by the ELST. The presented case of a VHL-associated ELST with increased  $^{68}\text{Ga}$  DOTATATE uptake indicates cell-surface expression of somatostatin receptors by this tumor, suggesting the potential application of somatostatin receptor imaging using  $^{68}\text{Ga}$  DOTA-conjugated peptides in the workup and management of these patients.

**Key Words:** endolymphatic sac tumors, von Hippel-Lindau disease,  $^{68}\text{Ga}$ -DOTATATE PET/CT, somatostatin receptors-imaging

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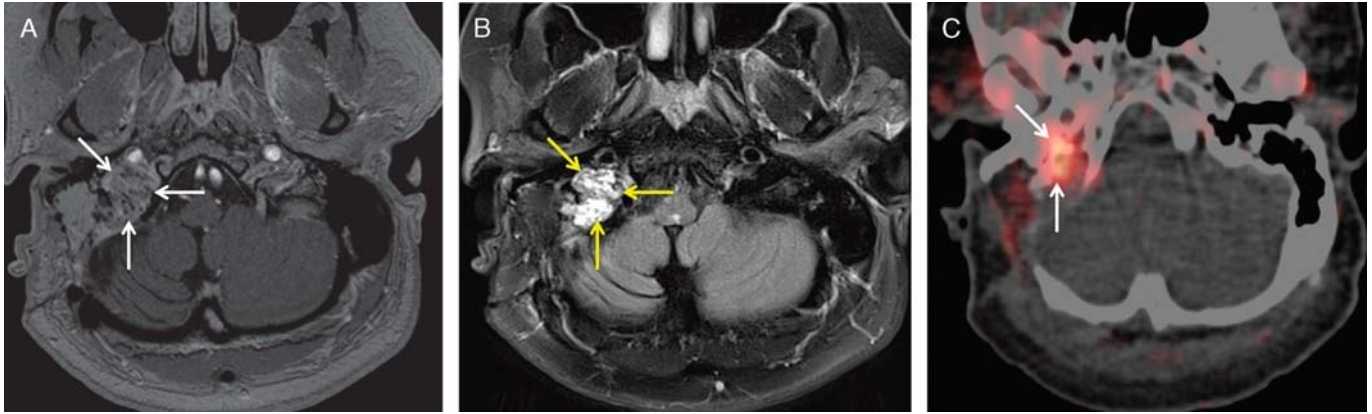
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**FIGURE 1.** A 46-year-old man with known history of von Hippel-Lindau (VHL) disease and partial resection of right-sided temporal bone mass histologically proven to be endolymphatic sac tumor (ELST) was followed up over a period of 14 years. Surveillance imaging with consecutive MR scans showed stable residual ELST localized within the right temporal bone causing destruction of all right inner ear structures (postcontrast T1-weighted image, white arrows, **A**), markedly enhancing on postcontrast FLAIR images (**B**, yellow arrows). In addition to anatomical imaging, the patient was evaluated with whole-body PET/CT using  $^{68}\text{Ga}$  DOTATATE for the detection of neuroendocrine lesions, showing increased activity ( $\text{SUV}_{\text{max}}$ , 6.29) by the residual ELST (**C**, white arrows). Endolymphatic sac tumors are rare, slow-growing, locally aggressive, low-grade papillary malignancies originating from the epithelium of the endolymphatic duct and sac.<sup>1</sup> Depending on tumor extension, ELSTs present with cochleovestibular dysfunction, sensorineural hearing loss, tinnitus, vertigo, ataxia, disequilibrium, or even facial weakness and glossopharyngeal nerve deficits.<sup>2</sup> Although ELSTs do not give distant hematogenous metastasis, “drop metastasis” to the spinal canal may occur, while death can happen secondary to intracranial vascular compromise.<sup>3,4</sup> Endolymphatic sac tumors can be either sporadic or associated with VHL disease, a familial cancer syndrome characterized by the development of a wide spectrum of benign and malignant tumors.<sup>5</sup> Von Hippel-Lindau–associated ELSTs appear to occur at younger ages, with predilection for women and are more frequently bilateral.<sup>6</sup> Microscopic abnormalities in the epithelium of the endolymphatic sac and duct similar to those of ELSTs are found in VHL patients even though they do not present clinical or radiological manifestations of the tumor.<sup>7</sup> Because multiple VHL manifestations such as hemangioblastomas or pancreatic neuroendocrine tumors are known to overexpress somatostatin receptors (SSTRs), SSTR imaging with radiolabeled SST analogs has been employed in the workup of this disease.<sup>8–10</sup> The conjunction of SST analogs with the chelator DOTA enabled labeling with the PET-emitter  $^{68}\text{Ga}$ , providing compounds with higher affinity to SSTRs compared with those used for conventional SSTR imaging with SPECT. Moreover, the superior resolution provided by hybrid PET/CT makes SSTR imaging using  $^{68}\text{Ga}$  DOTA-conjugated peptides the new standard of reference for the detection and characterization of SSTR-positive lesions.<sup>11,12</sup> The presented case of a VHL-associated ELST showing increased activity on  $^{68}\text{Ga}$  DOTATATE PET/CT suggests cell-surface overexpression of SSTRs and particularly SSTR-2 for which  $^{68}\text{Ga}$  DOTATATE has a predominant affinity. This observation sheds more light to the behavior of this rare tumor and indicates the potential application of PET/CT imaging with  $^{68}\text{Ga}$  DOTATATE in the early detection of ELST, which could enable less morbid operations with higher likelihood for cure and hearing preservation. Furthermore, monitoring of disease progression as well as theranostic application of peptide receptor radionuclide therapy in patients with residual or recurrent ELSTs are potential benefits of SSTR imaging using  $^{68}\text{Ga}$  DOTA-conjugated peptides that need to be further investigated.