

Accepted Manuscript

Title: Clinicopathological features of low-grade malignant endolymphatic sac tumors

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PII: S0344-0338(17)31102-0
DOI: <https://doi.org/10.1016/j.prp.2017.12.007>
Reference: PRP 51966

To appear in:

Received date: 2-11-2017
Revised date: 1-12-2017
Accepted date: 11-12-2017

Please cite this article as: Hong-Qun Wang, Li Jie, Huai-Yin Shi, Clinicopathological features of low-grade malignant endolymphatic sac tumors, Pathology - Research and Practice <https://doi.org/10.1016/j.prp.2017.12.007>

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Clinicopathological features of low-grade malignant endolymphatic sac tumors

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Abstract

BACKGROUND: Low-grade malignant endolymphatic sac tumor (ELST) is a rare neoplasm, occurring in the inner ear and invading the temporal bone. This study aims to investigate the clinicopathological features of low-grade malignant ELSTs. **METHODS:** The clinicopathological data of 21 patients with low-grade malignant ELSTs were collected and analyzed. **RESULTS:** The patients were aged 16-71 years, with an average age of 40.3 years and a median age of 39 years, and the male to female ratio was 1:1.6. There were 13 cases (61.9%) of ELSTs occurring on the left side, 7 cases (33.3%) on the right side, and 1 case (4.8%) on both sides. Blood types O and B were noted in 71.4% of the patients. Immunohistochemistry showed that CK, EMA and Vim were all positive, and S-100 (71.4%, 10/14), CD56 (75.0%, 9/12), NSE (50.0%, 2/4), and GFAP (11.1%, 1/9) were also positive, while Syn, CgA, TTF-1, TG, CD34, and calcitonin were negative. The Ki-67 index was 4.3% on average. Histologically, cells were arranged in a papillary shape often with branches and abundant fibrous axial vessel. Some cells had an expanded different-sized thyroid-follicle-like structure, with the follicles containing red-stained colloids and scallop-like secretory vacuoles. There were expanded cavities. Some cases were in a glandular arrangement, and a few in a nest-like, gland-cystoid arrangement. Most tumors were coated with a monolayer of cubic epithelium, a few cells were flat or columnar, with translucent cytoplasm and light staining. The nuclei were oval,

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