PHILIPPINE JOURNAL OF OTOLARYNGOLOGY-HEAD AND NECK SURGERY VOL. 32 NO. 1 JANUARY – JUNE 2017

Middle Ear Paraganglioma

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A 51-year-old woman underwent mastoidectomy with labyrinthectomy on the right for a polypoid external auditory canal mass accompanied by tinnitus and ear discharge. She was reported to have undergone mastoidectomy on the same site seven years prior to the present consult. The material from this prior surgery was not made available to us.

The submitted specimen from this surgery consisted of several dark brown irregular tissue fragments with an aggregate diameter of 4.2 centimeters. Histologic sections show tumor cells arranged in "ball-like" clusters that are surrounded by a network of sinusoidal channels. The cells are round to oval with round, uniform nuclei that have finely granular chromatin and moderate amounts of eosinophilic to amphophilic cytoplasm. (*Figure 1*) Mitoses, nuclear pleomorphism and hyperchromasia are not observed. Immunohistochemical studies show diffuse cytoplasmic positivity for synaptophysin and chromogranin. (*Figure 2*) The S100 stain highlights a peripheral layer of cells taking up the stain around the cell clusters. (*Figure 3*) Based on these features, we diagnosed the case as a paraganglioma likely a recurrence.

Paragangliomas are neuroendocrine neoplasms that arise from paraganglia found in various anatomic locations.[1] In the middle ear, they arise from paraganglia found in the adventitia of the jugular bulb – hence, the old synonym "glomus jugulare" and "glomus tympanicum." Other sites where they can develop include paraganglia of the carotid artery bifurcation ("chemodectoma"), the larynx and the vagal trunk ("glomus vagale"). The World Health Organization has simplified the nomenclature of these tumors by calling all of them simply "paraganglioma" and specifying the site involved.[1] In our case, it is likely a middle ear paraganglioma borne out by the history, clinical picture, and the morphology. Head and neck paragangliomas occur in adults from the 5th – 6th decade, more commonly in females, and present mostly with mass-related symptoms.[2,3]

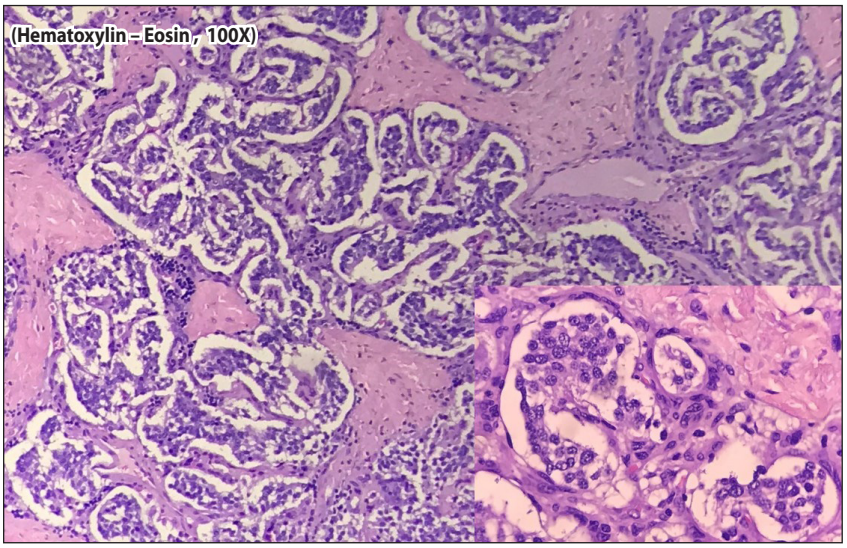

Figure 1. Tumor cells arranged in "ball-like" clusters surrounded by vascular channels (Hematoxylin-eosin, 100X magnification). High power (inset) shows round to oval cells with round uniform nuclei and finely granular chromatin (Hematoxylin-eosin, 400X magnification).

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Philipp J Otolaryngol Head Neck Surg 2017; 32 (1): 59-60

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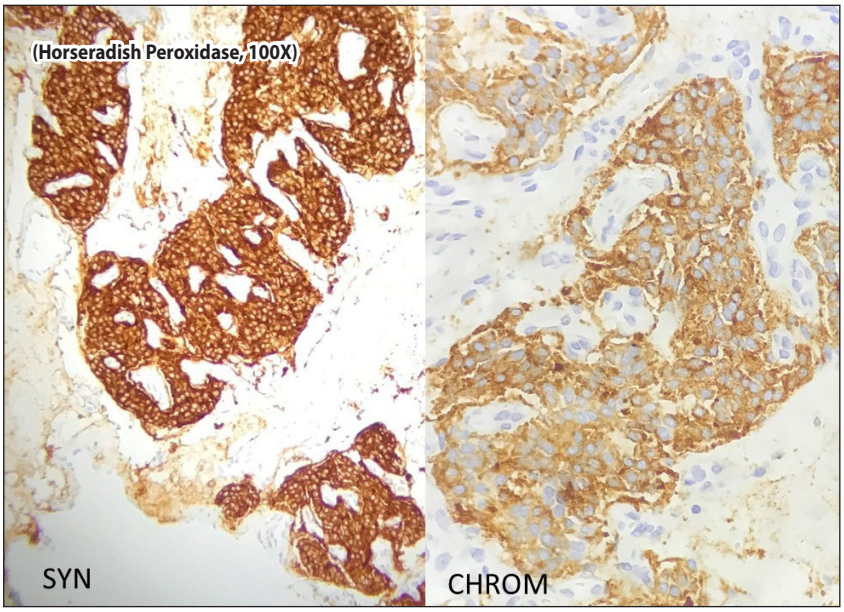

Figure 2. Diffuse cytoplasmic positivity on synaptophysin and chromogranin immunohistochemistry (Horseradish peroxidase method, 100X magnification).

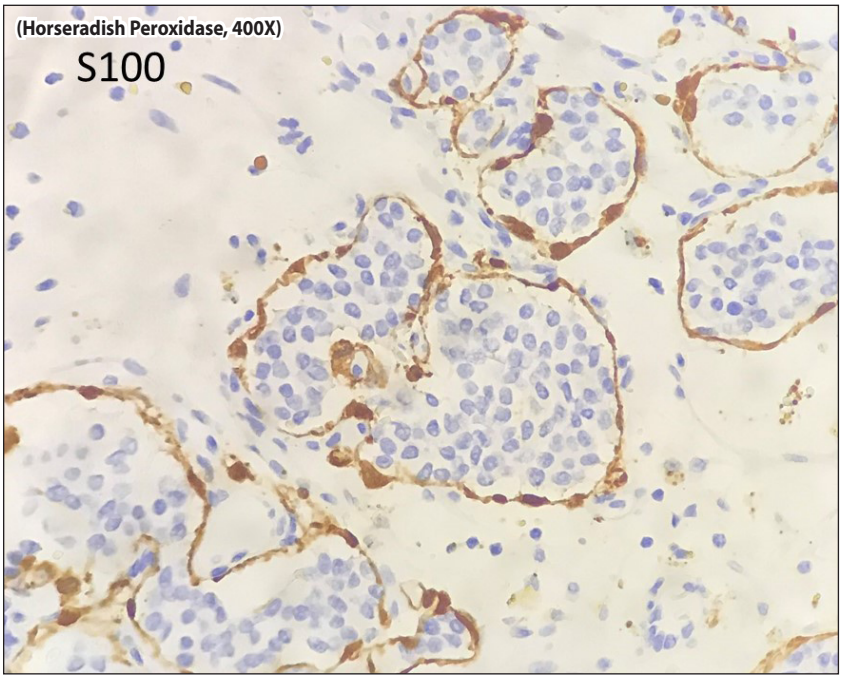

Figure 3. Peripheral positivity highlighting a layer of sustentacular cells around the cell clusters on S100 immunohistochemistry, (Horseradish peroxidase method, 400X magnification).

The morphology of paragangliomas in all head and neck locations is similar. Hematoxylin-eosin sections show cells arranged in organoid groups ("cell-ball", "Zellballen") surrounded by a vascular network. There are two cell types encountered: the chief cells which comprise the bulk of the cell nests and have abundant eosinophilic cytoplasm and the sustentacular cells which are spindly and located at the periphery of the nests. Neuroendocrine immunohistochemical stains (e.g. synaptophysin, chromogranin, CD56) highlight the chief cells while S100 and glial fibrillary acidic protein (GFAP) highlight the sustentacular cells. Cytokeratin is typically non-reactive and distinguishes this tumor from neuroendocrine tumors (i.e. carcinoid, neuroendocrine carcinoma) and middle ear adenoma.[1,3] There are no consistent histologic features that can discriminate between benign and malignant cases, nor are there criteria that can predict aggressive behavior and metastasis.[1,2,3]

Head and neck paragangliomas are slow-growing tumors and surgery is the most common treatment option. Radiotherapy is an option, especially for vagal paragangliomas where severe vagal nerve deficits occur in surgically treated cases.[1] Recurrence after surgery is reported to be less than 10% for carotid and up to 17% in laryngeal cases.[1] Metastasis on the other hand occur in 4 – 6 % of carotid, 2% of middle ear and laryngeal, and 16% of vagal tumors.[3] The World Health Organization nomenclature states that "all paragangliomas have some potential for metastasis (albeit variable)."[1] Thus, long-term follow-up may be prudent for all cases.

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