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Commentary

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William W. M. Lo[1]

It is unusual in this or perhaps in any medical journal to find a case report accompanied by a special commentary. Yet, in view of the strategic position occupied by the relatively rare endolymphatic sac tumor in our understanding of adenomatous tumors of the temporal bone and the recent reports of association between endolymphatic sac tumor and von Hippel-Lindau disease, an autosomal-dominant multisystem disorder, a commentary on the article by Meyer et al (1) is perhaps not only appropriate but also timely.

Although long under intense scrutiny for its role in Meniere disease (2, 3), the endolymphatic sac was not recognized as a source of neoplasms until 1984 when Hassard et al (4) serendipitously encountered a small vascular tumor in the sac during a decompression operation on a patient thought to have Meniere disease. Michaels (5) apparently had a similar experience. The tumors in both studies showed papillary cystadenomatous features like those of the tumor reported by Meyer et al (1).

As was the latter tumor, the tumor in Hassard's patient was thought initially to be a choroid plexus papilloma by the neuropathologists. Subsequently, Heffner (6), reexamining the material of the Armed Forces Institute of Pathology, found, on clinical, histologic, electron microscopic, and immunohistochemical grounds, 20 tumors of probable endolymphatic sac origin, including the one reported by Hassard et al. All were locally invasive papillary tumors involving the posterior petrous bone and the adjacent posterior cranial fossa and had, in the past, been variously classified pathologically as, for example, ceruminal neoplasm (five), choroid plexus papilloma (four), and metastatic papillary thyroid carcinoma (three).

For years, confusion reigned over the classification of adenomatous tumors of the temporal bone (7). Since the mid-1970s, however, two distinct entities have emerged: first, middle ear adenomatous tumors (8) of the mixed histologic pattern (solid, trabecular, and acinar) and, more than a decade later, adenomatous tumors of the papillary pattern (9). The former, originating from the middle ear, are, by and large, nondestructive and spare the facial nerve. The latter, originating from the posterior petrous bone, are locally destructive and frequently invade the facial nerve in the medial mastoid (10). Neither tumor has been reported to metastasize. The evidence is now convincing that tumors of the temporal bone of the papillary type in fact arise from the endolymphatic sac (1, 4–6, 11–15).

"Carcinoid" tumors are now classified as middle-ear adenomatous tumors (10). Ceruminous gland tumors originate from the external auditory canal (5). Choristomas in the middle ear consist of ectopic salivary gland tissue (16).

The recognition of endolymphatic sac tumor as an entity has helped to explain previously perplexing cases of "cerebellopontine angle ceruminous gland tumors" (17, 18), "extradural choroid plexus papillomas" (19, 20), and "metastatic papillary carcinomas of unknown origin" (6), and so forth. As in the patient of Meyer et al, modern imaging has helped to pinpoint papillary tumors to the endolymphatic sac region and has placed radiologists in a position to suggest the correct diagnosis (1, 15). Although endolymphatic sac tumors are rare, as their radiologic features become better known, more will undoubtedly be recognized (15) (Baum PA, Dillon WP. Imaging features of adenomatous neoplasms of the temporal bone. Presented at the 26th Annual Conference of the American Society of Head and Neck Radiology, Vancouver, BC, May 13–16, 1993).

Most of the reported endolymphatic sac tumors apparently have been sporadic. However, several papillary tumors of the endolymphatic sac in patients with von Hippel-Lindau disease

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have now been documented in the otolaryngologic literature (11–13, 18, 21). Some patients with von Hippel-Lindau disease have *bilateral* endolymphatic sac tumors (12, 13). Recently, two cases of endolymphatic sac tumor in von Hippel-Lindau disease patients have also been presented (Quallet JC, et al. Imaging of temporal bone vascular tumors in von Hippel Lindau disease. Presented at the 7th Annual Meeting of The European Society of Head and Neck Radiology, Sitges, Spain, May 2–5, 1993).

Although von Hippel-Lindau disease has been well studied by neurologists and radiologists (22, 23), the prevalence of endolymphatic sac tumor among patients with von Hippel-Lindau disease is still unknown. Some of these tumors may have been assumed to be hemangioblastomas or metastatic hypernephromas.

While generally nonlethal, endolymphatic sac tumor, as in the patient of Meyer et al, frequently causes significant disability by involvement of the facial nerve (1, 6, 9, 10). In cases of bilateral endolymphatic sac tumors, facial diplegia may result. Alertness on the part of the radiologist, when reviewing images of patients with von Hippel-Lindau disease, may lead to early detection and early resection of endolymphatic sac tumors before they cause facial palsy.

Other implications from this newly recognized association may also be considered. For example, are patients in their second or third decades who have developed an endolymphatic sac tumor suspects of von Hippel-Lindau disease? Should they undergo genetic studies and genetic counseling as suggested for patients with von Hippel-Lindau disease (23)? Should they be monitored for possible development of other manifestations of von Hippel-Lindau disease?

We have much more to learn.

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