The Journal of Laryngology and Otology
February 1997, Vol. **111**, pp. 197–198

Letters to the Editor

Laryngeal paragangliomas. A review and report of a single case

Dear Sir,

We read with interest the recent paper by Özünlü *et al.* (1996) entitled Laryngeal paraganglioma. A review and report of a single case that appeared in the June issue of *JLO*. Because of our long time interest in this subject, we feel compelled to respond to several issues contained in this report.

We had hoped that our recent manuscript (Ferlito *et al.*, 1994) would have resolved many of the problematic issues relative to laryngeal paraganglioma. Apparently it has not. The article by Özünlü *et al.* (1996) continues to perpetuate many of the erroneous concepts relative to laryngeal neuroendocrine tumours in general, and laryngeal paragangliomas, in specific. Özünlü and colleagues appear unsure as to how they wish to approach the general issues of laryngeal paraganglioma. On the one hand, they continue to propagate the myth, as proposed by El-Silimy and Harvy (1992), relative to an outdated classification that divides laryngeal paragangliomas into type I and II categories. According to this scheme, type I laryngeal paragangliomas are generally benign, while type II are more often malignant and characterized by chronic pain in the throat. This classification is no longer valid. Type I laryngeal paragangliomas are now recognized as being genuine paragangliomas whereas type II laryngeal paragangliomas are actually malignant tumours, particularly atypical carcinoids, that have been mistaken for paragangliomas. On the other hand, Özünlü and colleagues offer the data that clearly refute this concept as determined by Barnes (1991) and by Ferlito *et al.* (1994). However, in so doing their article is confusing, contradictory and outright erroneous. For example, Özünlü *et al.* (1996) state that the case reported by Olofsson *et al.* (1984) 'should be accepted as malignant laryngeal paraganglioma (subglottic) due to metastasis of cervical nodes'. We have contacted Professor Olofsson who has indicated to us in a letter written June 25, 1996, that this patient has no signs of metastases to the cervical lymph nodes. Professor Olofsson further informs us that after the patient underwent a total laryngectomy (the only therapy administered), she has been well and is still alive without signs of recurrence. This is 12 years after the publication of that case report and 19 years after total laryngectomy (at the time of the report in 1984, the patient had already been followed-up for seven years). Obviously, Özünlü and colleagues have misinterpreted the laryngeal paraganglioma reported by Olofsson *et al.* (1984) as being malignant.

Although the case of laryngeal paraganglioma described by Özünlü *et al.* (1996) may be authentic, the diagnosis was made solely on its light microscopic appearance and the results of an S-100 protein immunohistochemical stain! The Zellballen pattern of growth is not unique to paragangliomas. It can be seen in a variety of other tumours, such as typical and atypical carcinoids, malignant melanoma and medullary carcinoma of the thyroid, etc. The S-100 protein is also nonspecific. In order to eliminate any doubt about the authenticity of any alleged laryngeal paraganglioma reported in the future literature, we strongly urge that each case be subjected to rigorous light microscopic, immunohistochemical and ultrastructural evaluation. At the light microscopic level, laryngeal paragangliomas should composed of both chief (epithelioid) and sustentacular cells arranged in a Zellballen pattern. With immunocytochemistry, the chief cells should at least be positive for synaptophysin, chromogranin and/or neuron-specific enolase and negative for epithelial markers (cytokeratin, epithelial membrane antigen and carcinoembryonic antigen) and calcitonin. The sustentacular cells, in turn, must be positive for either S-100 protein or glial fibrillary acidic protein. Ultrastructurally, neurosecretory granules should be found in abundance. Lastly, the authors indicate that laryngeal paragangliomas contain epithelial mucin. They do not. The presence of any mucin immediately eliminates laryngeal paraganglioma from consideration.

We can only reiterate that which we have explicitly detailed previously (Ferlito *et al.*, 1994) including an extensive critical review of the literature, that laryngeal paragangliomas are benign neoplasms that can be treated conservatively but require complete surgical excision. The prognosis following complete surgical removal is excellent. Confusion with neuroendocrine carcinomas (i.e., atypical carcinoid tumours) has lead to incorrect diagnosis and inappropriate classification schemes erroneously suggesting that laryngeal paragangliomas have the potential for aggressive behaviour. It is our belief that malignant biologic behaviour associated with laryngeal paragangliomas is an extraordinarily rare occurrence (less than two per cent of cases) (Ferlito *et al.*, 1994). Conversely Özünlü and colleagues continue to quote old literature and propagate the myth that about 25 per cent of all laryngeal paragangliomas are malignant. Aggressive clinical behaviour of a presumed laryngeal paraganglioma

probably indicates that the tumour is not a paraganglioma but represents one of the classes of neuroendocrine carcinomas. Attention to the clinical behaviour as well as the light microscopic features and immunohistochemical antigenic profile should prevent incorrect diagnosis and classification.

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Polymorphous low-grade adenocarcinoma (PLGA) of the tongue

Dear Sir,

We read with interest the case report entitled 'Polymorphous low-grade adenocarcinoma of the tongue' by de Diego *et al.* (July 1996). We would like to share our experience with a case of PLGA arising from the glossotonsillar sulcus.

A 50-year-old female presented with history of foreign body sensation the right side of her throat for the past six months. There was no history of difficulty in swallowing or change in voice. On examination there was a 1.5 × 1.5 cm mucosa covered, firm mass on the right glossotonsillar sulcus with induration extending on to the right tonsil and adjacent base of the tongue. Biopsy under local anaesthesia was reported as low-grade adenocarcinoma. Chest X-ray, ultrasonography of the abdomen, thyroid scan (radioactive iodine^{131} uptake) and bone scan were normal. CT showed well demarcated mass on the right glossotonsillar sulcus with extension to the base of the tongue and the right tonsil. Subsequently the patient underwent wide excision of the tumour including partial mandibulectomy, neck dissection and reconstruction with pectoralis major myocutaneous flap. Post-operative recovery was uneventful. On gross examination there was a 3 × 2 × 2 cm greyish-tan mass. The final histopathological report was PLGA. Areas of prominent papillary pattern were noted. All the lymph nodes examined histopathologically were free of metastasis. No post-operative radiotherapy was given. Two years since her operation she remains asymptomatic and free of any evidence of recurrence.

The authors' comment on the role of prophylactic neck dissection in PLGA seems to be very much valid. Retrospectively we feel that in our case neck dissection could have been avoided. We are keeping our patient under regular close follow-up in view of worse clinical behaviour of PLGA with papillary elements.

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Erratum

Arytenoidopexy for bilateral vocal fold paralysis in young children. *JLO* **110 (11):** 1027–1031. Patients and Methods section read 'in a few cases there was intermittent abduction movement' this should have read 'in a few cases there was intermittent adduction movement'. We apologise for any inconvenience caused. ED.