GLOMUS INTRAVAGALE TUMOUR—A CASE REPORT

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Paragangliomas are uncommon tumours arising from chemoreceptor organs. Those arising in relation to the vagus nerve are referred to as Glomus intravagale or vagal body tumours. They are rare tumours accounting for only 3% of Head & Neck paragangliomas. A case of Glomus intravagale tumour is presented both in view of its rarity and to stress its differentiation from a carotid body tumour which in this case was possible only at the time of surgery.

Key words: Paraganglioma, Glomus intravagale, vagus nerve.

Case Report

A 30 year old male patient presented with history of a progressively enlarging mass in the right side of the neck for the past 3 years. A previous exploration of the mass elsewhere for suspected tuberculous lymphadenopathy was aborted after it was found that the mass was deeply situated and no biopsy performed. At presentation to us, patient had unilateral headache, discomfort while swallowing and some change in the voice. On examination, he had a 10x6 cms firm, mobile mass in the upper part of the right side of the neck extending from the level of the thyroid cartilage to the angle of the mandible. There were no transmitted pulsations and the mass was pushing the lateral pharyngeal wall resulting in a bulge posterior to the tonsil. The soft palate movements were normal, however, there was restriction in the mobility of the right vocal cord. A FNAC of the mass was inconclusive.

A limited CT done elsewhere which the patient brought with him was suggestive of a parapharyngeal tumour and the patient was explored through a cervical approach. At exploration, a vascular mass intimately related to the carotid vessels was encountered and an intra-operative diagnosis of a carotid body tumour was made. Since arrangements for a Vascular by-pass had not been made and enough blood for transfusion was not available, it was decided to close the wound and re-explore at a later date with a vascular surgeon available in case there was a need to resect and graft the internal carotid artery.

A more detailed CT examination of the neck carried out subsequently revealed a moderately enhancing mass extending up to the base of the skull. The carotid vessels were not separately identifiable from the mass (Fig. 1). In view of the patient's financial constraints and since findings at surgery were already suggestive of a
paraganglioma—possibly a carotid body tumour; patient was not subjected to an angiography. On re-exploration, to our surprise, the mass was found to be posterior to the carotid vessels indicating that this indeed was a glomus intravagale tumour! The mass was completely removed after sharply dissecting it from the vagus nerve and in the process some of the fibers of the nerve had to be sacrificed. Similarly, the XII nerve was also freed from the mass with sharp dissection.

Post-operatively, patient has weakness of the X and XII cranial nerves with deviation of the tongue and an immobile right vocal cord which are expected to partially recover. He is free from headache and is able to take a normal diet without aspiration of dysphagia and has satisfactory voice. The histopathology of the mass was consistent with a chemodectoma.

Discussion

Paragangliomas (syn. Glomus tumour, Chemodectoma, Non-chromaffin paraganglioma) are neoplasms that arise from paraganglial tissues, which are chemoreceptor organs distributed throughout the body. These organs are of neural crest origin and have similar function and possess similar histologic, ultrastructural and cytochemical features (Batsakis, 1979). Paragangliomas arising from the Ganglion Nodosum (Inferior ganglion of the vagus) and the cervical portion of the vagus nerve are referred to as glomus intravagale (Conley, 1977) or vagal body tumours (Kahn, 1976). They are rare tumours constituting only 3% of Head and Neck paragangliomas (Hirsch, 1982). They classically present as slow growing neck masses which cause paralysis of the vocal cord early in their clinical course (Leonetti, 1989). As they grow, they extend up to the skull base and may be intimately adherent to the internal carotid artery and other cranial nerves coming out of the skull base. Typically, the carotid vessels are pushed medially and anteriorly by the mass, unlike a carotid body tumour which pushes the internal carotid artery laterally and posteriorly, and causes splaying of the carotid bifurcation (Fig. 3).

In this particular case, although the presentation of a neck mass and a partially paralysed vocal cord should have alerted us to the diagnosis of a glomus intravagale tumour, the rarity of the condition and the fact that the authors had not come across a similar case earlier, resulted in the patient undergoing an unnecessary initial exploration. In addition, the lack of identification of the carotid vessels separately from the mass on CT Scan