Introduction

THE VAGAL paraganglioma (VP) arises from paraganglionic tissue located along the vagus nerve. This neoplasm represents less than 5% of all head and neck paragangliomas, and since its original description in 1935, fewer than 200 cases have been reported in the literature. It arises from an island of paraganglionic tissue derived from the neural crest that is located on the vagus nerve. The growth of these tumors is very variable, some grow upward toward the skull base while others extend downward into the neck. Because of their rarity, most of the information about these tumors is from case reports and small series. In many cases, they are “lumped” together with other vagal nerve tumors or intermixed with other head and neck paragangliomas. Although many authors have advocated surgical excision as the primary modality of therapy for paragangliomas of the head and neck, the relative effectiveness of surgery vs radiation therapy remains controversial. Literature can be cited to support either modality. Radiation therapy, while it may significantly slow tumor growth and provide palliation in selected patients, has not been shown to cure paragangliomas. In most cases, the tumor size decreases only if it is irradiated and histologically viable tumor cells (chief cells) have been shown to be present after radiation therapy.

Case summary

A 68-year-old male presented with a 4-months history of a neck mass. He had suffered a concurrent sporadic dry cough unrelated to eating, talking, or any particular movement. He reported concomitant intermittent hoarseness. He denied dysphagia; he had no family history of paragangliomas or other neuroendocrine tumors, or any relevant past medical history.

On examination, a right side 6x7cm neck mass deep to the sternocleidomastoid was present, which was discrete, mobile, and nonpulsatile. Palpation of the mass immediately triggered coughing. A blood panel was normal and 24-hour urine was negative for catecholamines. CT angiography of the neck demonstrated a heterogeneously enhanced irregular 4.4 x 4.2 cm mass within the right carotid sheath along the lateral aspect of the proximal internal and external carotid arteries with minimal spaying of these arteries. The central area of the mass was of lower attenuation suggestive of necrosis. Differential diagnoses included vagal paraganglioma, schwannoma, neurofibroma, or a necrotic lymph node. The patient was counseled on management options. Due to the impact of the tumor on his quality of life, he elected for surgical removal of the mass. The mass was excised through a 11 cm incision made two finger-breadths below the mandible, overlying the sternocleidomastoid. Right pericardial and lymph nodes were resected to improve access to the mass. The mass was dissected free from the internal and external carotid arteries and the internal jugular vein. The vagal nerve was intimately involved with the mass. The spinal accessory and hypoglossal nerves were identified and preserved. He tolerated the operation well and discharged two days post operative. He developed post operative chocking for about three weeks then improved.

The pathology report described a completely excised encapsulated 4x3x1.5 cm. it is firm, with greyish cut surface. Examination revealed a fairly capsulated tumor mass. Neoplastic cells are arranged in nesting pattern, with intervening vascular spaces (zellballen pattern). Neoplastic cells are polygonal with moderate amount of cytoplasm, and occasional vacuolation (lipoblast-like). These exhibit moderate degree of pleomorphism with scanty mitoses. So the patient referred to oncology medicine for follow up management.

Discussion

Paragangliomas arise from the paraganglia, which are small groups of neuroendocrine cells stemming from autonomic nervous system ganglia. Usually slow growing and benign, tumors of the paraganglia are most common in the adrenal medulla (pheochromocytoma), with 85% of extra-adrenal PCs in the abdomen, 12% in the thorax, and 3% in the head and neck. Four genetic PG syndromes have been described, all with autosomal dominant transmission. Surgical excision is the classical treatment of choice for most VPs; however, contemporary management is evolving toward more conservative measures due to the high associated morbidity. Treatment planning for these patients can be fraught with problems and the major dilemma is the choice between surgery and observation. Radiotherapy as a realistic alternative lacks an evidence base. The decision must be based on several factors that include the age of the patient, the preoperative status of the vagus, and the size and growth rate of the tumor. Multifocal disease may modify the management plan radically. In this respect there are some basic facts that should be considered.

The natural morbidity of glossopharyngeal and vagal deficits is better tolerated than that caused by surgery. The elderly do not compensate well for glossopharyngeal and vagal deficits. Large tumors may inflict other cranial nerve deficits. Small tumors, which we consider to be equal to or smaller than 2 cm in diameter, are usually easily removed with a predictable morbidity that is limited to the vagus nerve only, and may well be a preferable deficit in any case. A large tumor carries with it higher surgical risks with the possibility of additional cranial nerve palsies and damage to the internal carotid artery. Modern imaging techniques allow accurate monitoring of tumor growth that allows a change of management plan before the risk of collateral damage becomes too great.

Conclusion

Vagal paragangliomas usually arise in the parapharyngeal space though there is evidence of cervical paragangliomas growing along the cervical course of the vagus. The cervicotoparotid approach using microsurgical techniques gives safe access to the upper pole of the tumor with minimal risk to the lower cranial nerves and the internal carotid artery. Management options for patients with these tumors include watchful waiting and surgery. The choice between these options is determined by consideration of the likely natural history, age of the patient, size of the tumor, and associated cranial nerve palsies. Surgical treatment of the vagal tumors in patients with multifocal disease should only be undertaken if it will not add to the patient’s morbidity. Progress in molecular biology and genetics should help in the screening and treatment planning of familial cases.

Fig 1: tumour in relation to carotid arteries
Fig 2: After Tumour excision preserved cranial nerves
Fig 3: Usual carotid art. exposure
Fig 4: Tumour size after excision