

MALIGNANT PARAGANGLIOMA OF THE HEAD AND NECK. WHAT DO WE KNOW?

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Introduction

- Malignant paragangliomas (MPG) are exceptionally rare tumors corresponding to 0.016-19% of all head and neck paragangliomas (HNPG). Unlike other tumors, MPGs are diagnosed when there is pathologically confirmed paraganglioma cells in lymph nodes or in distant organs, rather than by specific criteria found on pathologic analysis. In this report we present two patients with MPGs who illustrate the varied presentation of this rare entity.

Case 1

- 36-year-old female with a 3-year history of a right neck mass. Her MRI revealed a right 4.7 cm mass inferior to the carotid bifurcation (CB) with an adjacent ipsilateral 3 cm satellite lesion and a similar, 2.7 cm mass centered on the left CB (Figure 1).

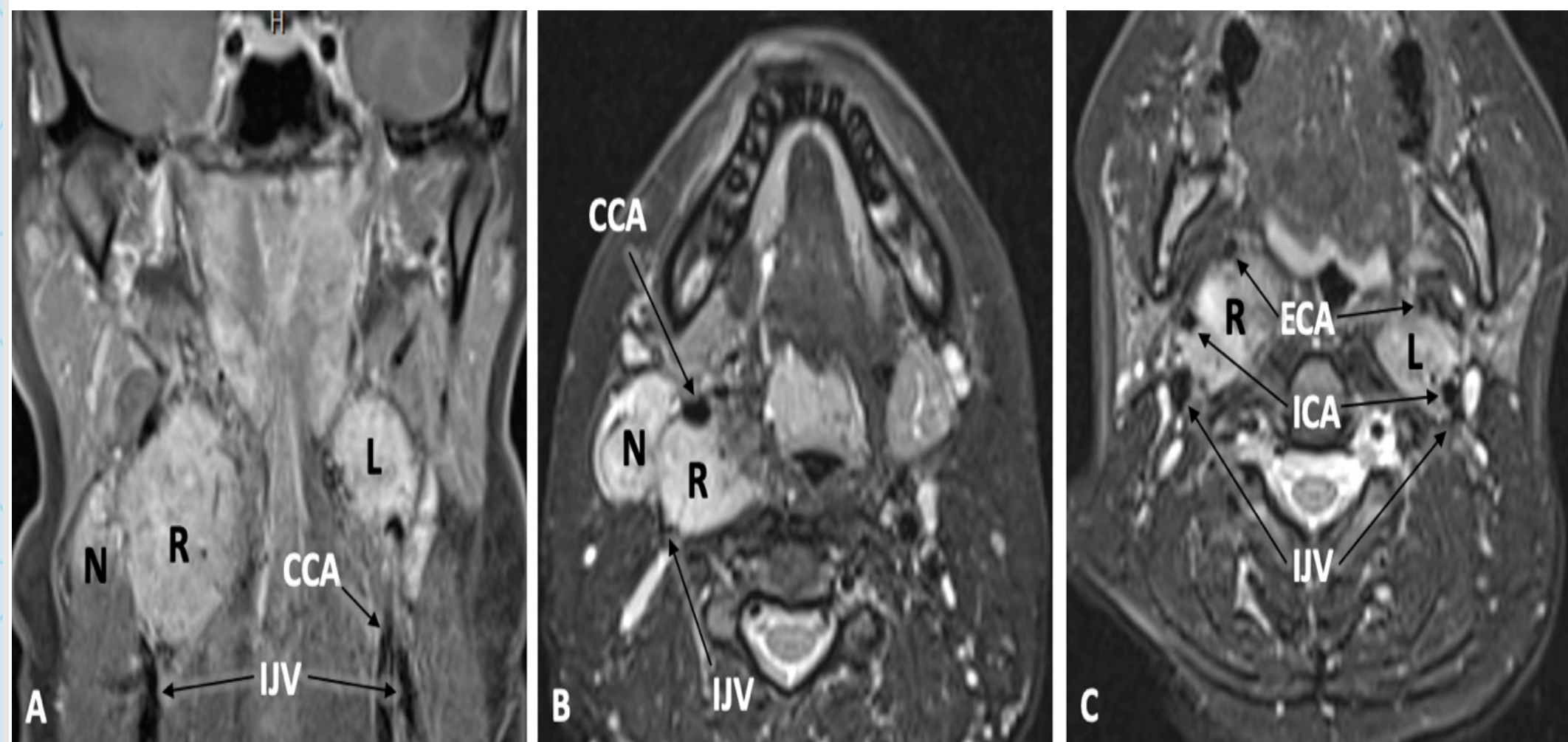


Figure 1. Preoperative MRI of Patient 1. A) Image of bilateral paragangliomas and right involved lymph node. B) Image of right 4.7 cm paraganglioma with adjacent, 3 cm node and displacement of CCA and IJV posteriorly. C) Left, 2.7 cm paraganglioma splaying the ECA and ICA and posteriorly displacing the IJV. Additionally, the right tumor is also visualized and its relation to the ECA, ICA and IJV. R, right paraganglioma; L, left paraganglioma; N, lymph node; CCA, common carotid artery; ECA, external carotid artery; ICA internal carotid artery; IJV, internal jugular vein.

- Underwent preoperative angiography, balloon occlusion testing and tumor embolization and resection of only right sided tumors with pathologic diagnosis of paraganglioma (larger tumor) with positive margins and regional nodal metastasis to a lymph node (smaller tumor) with lymphovascular invasion (Figure 2). She received primary RT and adjuvant RT for left and right neck, respectively, with post-RT PET scan revealing unchanged left paraganglioma, bilateral metastatic retropharyngeal nodes (larger on the right) and a metastatic frontal bone lesion. Her follow up scans have demonstrated stable local, regional and distant disease and patient is currently undergoing mAb treatment for bone metastasis.

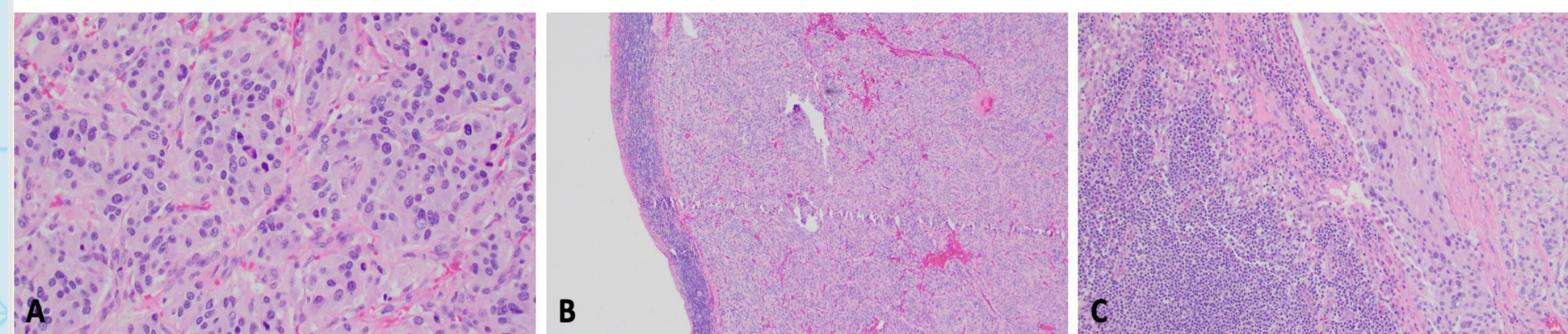


Figure 2. Micrographs of pathologic analysis with hematoxylin and eosin stain of patient 1 representing cytomorphologic characteristics of paraganglioma (A), nodal metastases (B) and lymphovascular invasion of tumor (C).

Case 2

- 27-year-old female with a 3-month history of unilateral submandibular discomfort with a magnetic resonance (MRI) revealing a 6cm, suprahyoid neck mass encasing the internal (ICA) and external carotid artery with features suggesting a carotid body tumor (CBT), and no pathologically enlarged lymph nodes (Figure 3). She underwent preoperative angiography, balloon occlusion testing and tumor embolization, unilateral tumor resection and concurrent levels II-IV neck dissection (Figure 4), with a pathologic diagnosis of paraganglioma and a tumoral deposit found in one lymph node. She is currently under surveillance given clear margins. Postoperative imaging demonstrated residual tumor within carotid space and pulmonary metastatic disease. She is currently undergoing evaluation by radiation oncology for adjuvant radiation therapy.

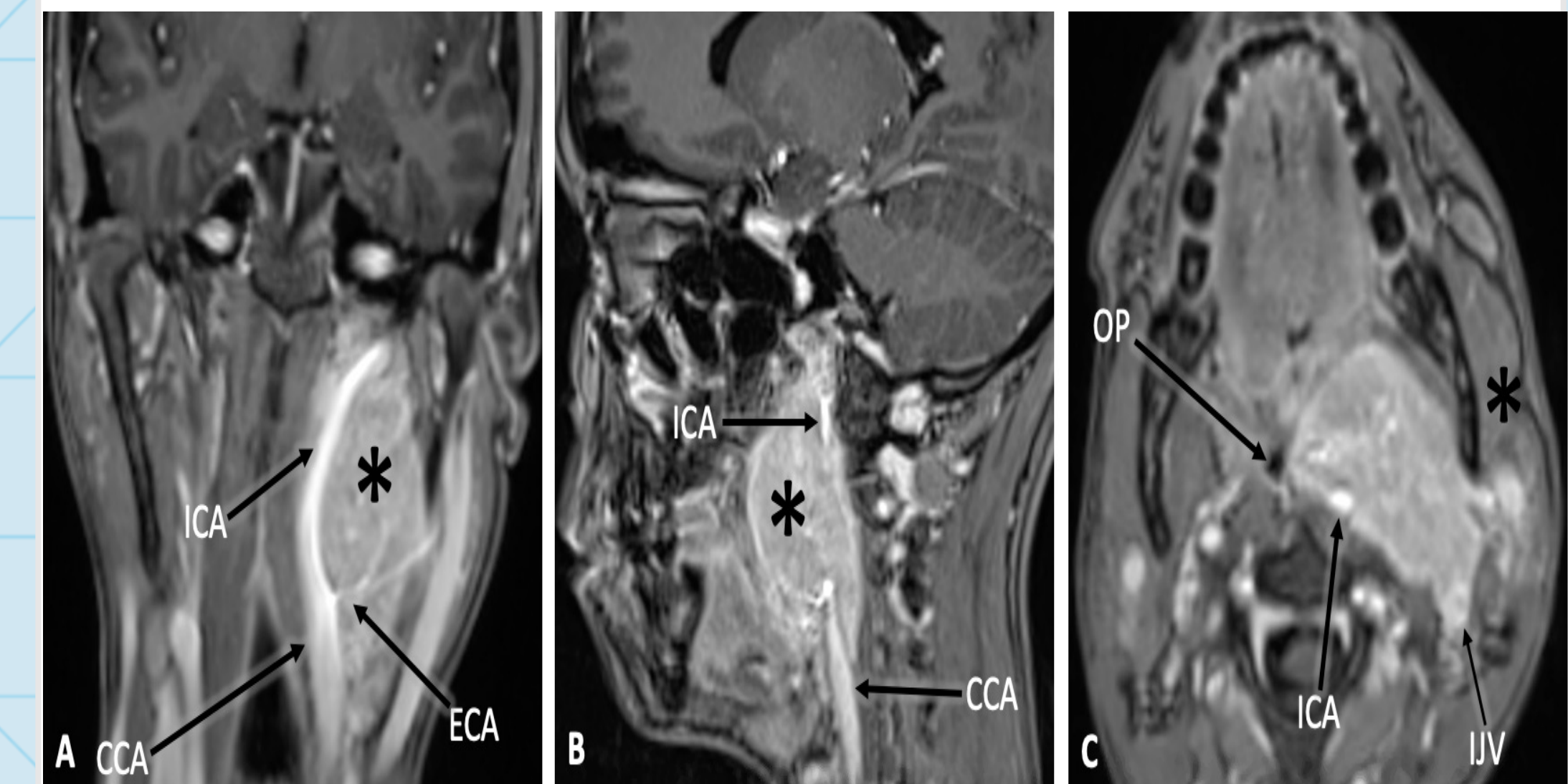


Figure 3. Preoperative MRI of Patient 2 demonstrating 6 cm, suprahyoid paraganglioma, involving the carotid and parapharyngeal spaces, encasing the ECA (A) and ICA (A, B, C) with notable narrowing of the upper aerodigestive tract (C). *, paraganglioma; CCA, common carotid artery; ECA, external carotid artery; ICA, internal carotid artery; IJV, internal jugular vein; OP, oropharynx.

Summary

MPGs are exceedingly rare head and neck tumors and represent a small fraction of all HNPG. Vagal HNPGs are the most likely to be MPG, but given the higher occurrence of CBTs, these are more commonly seen in a malignant form. The hereditary form is more likely to harbor malignancy. The diagnosis of MPG is made when there is evidence of nodal involvement and/or presence of metastases to nonendocrine tissue. For any HNPG, surgical resection with neck dissection is necessary to determine presence of nodal metastasis and prevent delay in potential adjuvant treatment. Given the rarity of MPGs, adjuvant chemotherapeutic and radiation strategies continue to evolve given the unclear effect on survival and unclear benefit following surgery. Given the malignant nature of these tumors and association of paragangliomas to genetic syndromes, long term follow up and surveillance for regional and distant recurrence and presence of tumors in other parts of the body is recommended.

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