Paratesticular Paraganglioma: A Case Report and Overview of Imaging Modalities

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Learning objectives

- Present a case of a paratesticular paraganglioma.
- Review the imaging modalities used in the diagnosis and staging of paragangliomas.
- Discuss imaging characteristics of different types of paragangliomas.
Paragangliomas (PGLs) are tumours arising in crests of neuroendocrine cells (paraganglia), that are classified by both location and secretory function. Parasympathetic PGLs are typically located in the head & neck and present with mass effect; sympathetic PGLs occur along the sympathetic chain and present with catecholamine excess. By definition, phaeochromocytomas are intra-adrenal sympathetic PGLs.

PGLs are the largest group of hereditary tumours, with mutations commonly in the succinate dehydrogenase subunit (SDHx).\(^1\) Associated hereditary syndromes include von Hippel-Lindau (VHL), multiple endocrine neoplasia types 2A (MEN2A) and 2B (MEN2B), neurofibromatosis type 1 (NF1) and Charney-Stratakis dyad. SDH-D mutations are common in head and neck PGLs (HNPs). SHD-B mutations are typically associated with sympathetic PGLs and confer a higher risk of malignancy.

Malignancy is defined as evidence of metastases, with the most common sites being lymph nodes, lung, liver and bone. Approximately 10% of phaeochromocytomas, 20% sympathetic PGLs and 2-20% of parasympathetic PGLs are malignant.
Imaging findings OR Procedure details

Case Report: A 55-year-old male presented with a one-year history of hoarse voice, and was found to have a right vocal cord palsy on examination. CT neck demonstrated a well-defined homogenously enhancing lesion in the right carotid space with punctate foci of vascular enhancement (Figure 1 and 2). On MRI the lesion appeared hypointense and heterogenous on T1 and T2 imaging with mild post-contrast enhancement (Figures 3-5). Characteristic 'salt and pepper' morphology was present, suggesting a diagnosis of a paraganglioma, likely of vagal origin, with the differential of a Schwannoma.

Further tumour characterisation with $^{18}$Fluro-2-deoxyglucose ($^{18}$F-FDG) positron emission tomography (PET) demonstrated an FDG-avid lesion in the right nasopharyngeal region (Figure 6). Unexpectedly, abnormal uptake was also seen in the left hemiscrotum (Figure 7). On clinical examination, there was tender thickening of the left spermatic cord with normal left epididymis and testis. On Ultrasound, the abnormality corresponded to a 21x21x15mm ovoid solid low echogenicity mass arising within the right spermatic cord with marked internal vascularity. This lesion was noted to be separate from the normal left testis and epididymis. Following surgical excision, the lesion was histologically proven to be a paraganglioma. Genetic testing showed an SDHB mutation.

Overview of Imaging Modalities: Imaging of PGLs requires a combination of anatomical and functional modalities. Anatomical imaging involves CT and MRI. Functional imaging includes several nuclear medicine tracers, with varied utility depending on tumour characteristics.

Anatomical Imaging

CT and MRI are often the initial imaging modalities for tumour localisation. They have excellent sensitivity but lack specificity in unequivocally identifying a mass as a PGL; however, characteristic findings can strongly suggest a diagnosis. On unenhanced CT, PGLs typically appear as a mass with density greater than 10 Housefield units. With contrast, they avidly enhance due to a rich capillary network, with a delayed washout phase. On MRI T2-weighted images, PGLs typically have a hyperintense 'light bulb' appearance, although this can be variable with some lesions appearing hypointense, while on T1-weighted images, they generally have low to intermediate signal intensity. PGLs also have a characteristic 'salt-and-pepper' appearance due to enhancing parenchyma (salt) and signal flow voids of vessels (pepper). With gadolinium administration, there is heterogeneous, prolonged enhancement on T1 images.

Functional Imaging
Iodine-metaiodobenzylguanidine (\(^{123}\)I-MIBG) scintigraphy was the first functional modality used in PGLs and is highly sensitive in non-metastatic sympathetic PGLs. However, this technique, has limitations in head & neck PGLs (HNPs) and malignant disease.\(^2,3\) \(^{18}\)F-3,4-dihydroxyphenlyalanine (\(^{18}\)F-FDOPA) PET has proven superiority in detecting non-metastatic HNP, particularly SDHD mutations. It also performs well in metastatic disease, unless there is an underlying SDHB mutation, in which case there is a significant decrease in sensitivity (94% vs 20%, respectively).\(^2,4,5\) Conversely \(^{18}\)F-FDG is highly sensitive in metastatic disease, particularly SDHB-related cases.\(^6,7\) It also performs well in non-metastatic disease and has a similar specificity to \(^{123}\)I-MIBG.\(^6,8\) Gallium-DOTATATE PET is becoming increasingly recognised as a viable imaging modality for both hereditary and sporadic disease, and HNPs, performing better than all other modalities in lesion detection.\(^8-10\) \(^{18}\)F-flurodopamine is not readily available, but appears to have a role in metastatic disease, when compared to \(^{123}\)I-MIBG and \(^{18}\)F-DOPA.\(^2\)

**Paratesticular Paragangliomas**

Paratesticular PGLs are rare, with only twelve cases reported. The pathogenesis is unclear, although it has been suggested they arise from embryonic paraganglionic nests that descend with the testis during embryogenesis, rather than metastasis.\(^11\) Information on radiological characteristics is limited. They have been detected on \(^{123}\)I-MIBG in one instance, and now twice on \(^{18}\)F-FDG PET. Clinically they seem to present as a painless scrotal mass and without symptoms of catecholamine excess.
Fig. 1: Sagittal CT Neck demonstrating an intensely and homogeneously enhancing mass in the carotid space, with punctate foci of vascular enhancement.

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**Fig. 2:** Axial CT Neck demonstrating an intensely and homogeneously enhancing mass in the carotid space, with punctate foci of vascular enhancement.

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**Fig. 3:** Axial MRI T1 demonstrating a hypointense heterogeneous mass in the carotid space.

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Fig. 4: MRI T1 coronal post-contrast FS demonstrating a carotid mass with mild post-contrast enhancement, and typical 'salt & pepper' appearance.

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**Fig. 5**: MRI T2 sagittal neck demonstrating a hypointense and heterogenous mass in the carotid space.

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Fig. 6: 18F-FDG PET Head and Neck demonstrating an intensely FDG-avid mass in the right carotid space.

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**Fig. 7:** 18F-FDG PET Scan Whole Body demonstrating unexpected FDG-uptake in the right testicle.

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Conclusion

PGLs represent a group of tumours with various functional imaging characteristics depending on their location, secretory function and underlying genetic mutation. With now twelve reported cases, paratesticular paragangliomas could be considered a very rare differential for a scrotal mass. However, further understanding of their pathogenesis, association with genetic syndromes, and behaviour on functional imaging is required.
Personal information

Rachael is a medical registrar at the Townsville Hospital with a particular interest in Oncology and Haematology. Timothy is currently half way through his internship, also at the Townsville Hospital, with an interest in diagnostic radiology.
References