Paraganglioma Imaging: Are we adhering to International Guidelines? 
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Abstract

Purpose
Head and neck paragangliomas comprise 3% of extra-adrenocortical phaeochromocytomas. The aim of this audit is to define the current practice of paraganglioma (PGL) management amongst patients in the North West region referred to the Skull Base MDT.

Methods
A five-year retrospective audit was undertaken, investigating all patients referred through the Manchester Regional Skull Base MDT. Demographic data, diagnostic data and follow-up were collected from clinical notes using Sunrise and Chromeleon. Imaging methods including pre-diagnostic, diagnostic, renal and body imaging were analysed using PACS and CDSS.

Results
67 patients were included in the study, of which 75% were female. The majority of patients were new referrals. The most common primary diagnosis was glomus jugulare (77%), followed by glomus tympanicum (23%), 65% of patients referred to a clinical geneticist and 63% of patients undergoing genetic testing were found to have mutations of the SDH gene. MRI with contrast was most commonly used to diagnose paragangliomas. We found considerable heterogeneity in body imaging modalities utilised.

Conclusions
While most patients with suspected paragangliomas in the Greater Manchester Region are managed according to international guidance, there is considerable heterogeneity in body imaging performed. We plan to re-audit following implementation of a new paraganglioma Multidisciplinary Team including proposed standardisation of imaging protocols in the Manchester Skull Base Unit.

Introduction

1. Phaeochromocytoma
2. Paraganglioma
3. Paraganglioma: are neuroendocrine neoplasms of high vascularity, derived from the neural crest of the autonomic nervous system.

Paragangliomas

- Head & Neck PGLs
  - Mostly benign but have malignant potential
  - 6% demonstrate neurosecretory activity
  - Sporadic or 10-50 familial syndromes:
    - Multiple endocrine neoplasia type 2 – RET gene
    - Neurofibromatosis type 1 – NF-1 gene
    - Von Hippel-Lindau syndrome – VHN gene
    - Hereditary PGL syndrome – SDHB – SDHC/SDHD genes

- Paragangliomas
  - 85% intrabdominal
  - 15% head and neck
  - Glomus jugulare
  - Glomus vagale
  - Glomus tympanicum
  - Glomus vagale
  - Carotid body

- Phaeochromocytoma
  - 80-85% of patients referred for gene testing and to endocrinology
  - 48% patients referred for genetic testing

- Symptomatology
  - Otorrhoea
  - Rhinorrhoea
  - Otalgia
  - Hearing
  - Secretory PGL in 3 patients (10%)

- Diagnosis
  - Incidental: 24%
  - Clinical: 52%
  - Screening: 24%

- Imaging
  - MIBG-123
  - FDG-PET
  - MR

- Recommendations
  - FDG-PET
  - MR

Guideline

FDG-PET

MRI

MRI

MRI

MRI

FDG-PET

FDG-PET

Reference

