Secretory Head and Neck Neuroendocrine Tumour – A rare entity

Tamar Saeed¹, Michael Tadman¹, Aparna Pal¹, Samuel McKeith², Bahram Jafar-Mohammadi¹

¹Oxford Centre for Diabetes, Endocrinology and Metabolism (OCDEM), ²ENT department, Oxford University NHS foundation trust

Introduction: Paragangliomas can derive from either parasympathetic or sympathetic paraganglia; the two types occur with similar frequency³. The majority of parasympathetic ganglia-derived paragangliomas are located in the neck and skull base along the branches of the glossopharyngeal and vagus nerves³,⁴,⁵. They arise most commonly from the carotid body, less commonly from jugulotympanic and vagal paraganglia, and rarely, from the laryngeal paraganglia. The majority of paragangliomas arising within the skull base and neck region are not associated with catecholamine secretion; in various reports, up to 5 percent are symptomatic from hypersecretion⁶,⁷.

Presentation: We report a case of a 43 year old gentleman who had surgery in 1998 for left glomus tympanicum, leaving him with complete left lower facial nerve palsy and headaches. It is noted that he had labile blood pressure during surgery. There was gradual growth of the residual tumour associated with intermittent sweating.

In February 2016 he was referred to ENT for evaluation of his symptoms. In view of elevated metanephrines (Table 1) he was subsequently referred to endocrine service. In October 2016 he was confirmed to have a pathogenic mutation in succinate dehydrogenase B (SDHB) gene [heterozygous for SDHB c.311delinsGG, P.(Asn104fs)]. A likely indolent left level I Ib nodal metastasis was identified on FDG PET scan (Fig 1). The iodine-123-meta-iodobenzylguanidine (MIBG) scan demonstrated no increase uptake in the lesion (Fig 2). However, a skull base mass lesion and lymph nodes were avid on 68-Ga-DOTATATE PET CT(Fig 3).

Treatment: The patient was symptomatic and the biochemical markers confirmed the lesion to be functional (no other lesions were identified on scans). The utility of the Ga-68 DOTATE PET scan has provided a treatment option of Lutetium based Peptide Receptor Radionuclide Therapy (PRRT).

Conclusion: This case highlights that, though rare, head and neck paragangliomas can be secretory. Furthermore, the utilisation of appropriate functional imaging can be quite important in the treatment pathway. Genetic testing was carried out relatively late during the course of the management of this patient which may have given important information about likely course of the disease as well as imaging modalities that may have been useful in the detection of the disease.

References:
1. Pathology and Genetics of Tumours of the Endocrine Organs. WHO Classification of Tumours, DeLellis RA et al. 2004.

Table 1: Plasma Metanephrines – August 2016

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
<th>Reference Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>3 Methoxytyramine</td>
<td>1061 pmol/L</td>
<td>0-180 pmol/L</td>
</tr>
<tr>
<td>Metadrenaline</td>
<td>259 pmol/L</td>
<td>80-510 pmol/L</td>
</tr>
<tr>
<td>Normetanephrine</td>
<td>1888 pmol/L</td>
<td>120-1180 pmol/L</td>
</tr>
</tbody>
</table>

Fig 1: FDG PET Scan
Fig 2: MIBG
Fig 3: DOTATATE PET CT