

**A case of metastatic paraganglioma presenting 20 years after initial surgery**

*Lalantha Leelarathna<sup>1</sup>, Anna Crown<sup>2</sup>, Simon Aylwin<sup>3</sup>, Klaus-Martin Schulte<sup>4</sup>*  
*<sup>1,2</sup>Royal Sussex County Hospital, Brighton. <sup>3,4</sup>Kings College Hospital, London.*

**History:**

A 65 year old man was admitted to hospital in Dec 2006 with right sided pneumonia. He was on 4 anti-hypertensives with poor blood pressure (BP) control and had a history of excessive sweating and palpitations for many years. About 20 years ago he had developed a hoarse voice, pain and wasting of his left shoulder, with impaired balance and an abnormal gait. An MRI scan had revealed a left jugular foramen tumour and he had undergone surgery at Queens Square (QS). Borderline elevation of VMA excretion was noted post-operatively, but repeat VMA estimates in June 1989 were within normal limits and no further follow-up was arranged. His brother also has hypertension, and there is a history of early heart disease in his father and paternal grandfather. This extensive and important past medical history was not available in the patient's GP e-records or current hospital notes, and was obtained when his GP retrieved his written records which included correspondence from QS.

**Examination:**

Weakness and wasting of left trapezius together with paralysis of left 9, 11,12th cranial nerves were noted. His BP was 210/110 mmHg without postural drop. Pulse was 108 bpm, regular.

**Investigations:**

24 hour urinary catecholamines were markedly raised. Noradrenaline 27915 (NR <500) Adrenaline 68 (NR <100) and Dopamine 21164(NR <3000). CT scan of his abdomen showed a 12 cm mass arising from L.adrenal gland and multiple para-aortic and peri-caval lymphadenopathy. <sup>123</sup>I MIBG scan showed multiple MIBG avid foci extending from head to pelvis (base of skull, right supra clavicular fossa, posterior mediastinum, para spinal and para aortic regions) with a large lesion in the left suprarenal area with central necrosis. MRI brain showed no evidence of recurrence at the previous operation site.

**Management**

Alpha blockade was commenced. The patient was referred to the NET MDT at Kings College Hospital (Dr Simon Aylwin). On 04/04/2007 he underwent radical left adrenalectomy, left nephrectomy and dissection of sympathetic chain (Mr Schulte). Histology confirmed malignant phaeochromocytoma with lymph node metastases.

**Progress, further investigations and Follow-up**

Post operatively, urinary catecholamines levels have reduced by 25% but the patient has developed problems of low mood and postural hypotension, currently being treated with Fluoxetine and Fludrocortisone. He continues to suffer from episodic sweating and his lying BP is still raised. A recent MRI spine has shown extensive metastatic deposits throughout his lumbar and lower thoracic spine. Results of a repeat MIBG scan, MRI scan of abdomen and pelvis, and genetic analysis for SDHD / SDHB mutation are awaited. <sup>131</sup>I labelled MIBG therapy is being considered as the next therapeutic option.

**Discussion**

This case highlights the importance of life long follow-up of these patients who may present many years later with life-threatening consequences of their disease. In retrospect the diagnosis should have been considered earlier in view of his resistant hypertension and sweating. Recent studies have demonstrated a high incidence of genetic mutations in patients with phaeochromocytoma / paraganglioma indicating a need for more widespread genetic testing with

implications for family members. Management of these patients is complex and they should be referred to centres of excellence for early multidisciplinary input. Tumour debulking remains an important step and facilitates subsequent MIBG targeted therapy / combination chemotherapy.