



Large paraganglioma at the organ of Zuckerkandl

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A 34-year-old female with no prior medical history presented to gynaecology with lower abdominal pain and abnormal uterine bleeding. A suprapubic mass, believed to be a multifibroid uterus, was palpable on abdominal examination.

She was, however, noted to be both hypertensive (185/127 mmHg) and tachycardic (130 beats per minute) and had reported episodes of headaches and palpitations. She was referred to Internal Medicine for further workup.

Investigations revealed an elevated 24-hour urine normetanephrine (35,580 nmol; normal 480–2424) with 24-hour urine metanephrine within the normal range (369 nmol; normal 264–1729).

Magnetic resonance imaging (Fig. 1a) confirmed the presence of a large heterogeneous extra-uterine mass (11.7 × 10.8 × 11.7 cm) situated at the bifurcation of the aorta. An iodine-123 meta-iodobenzylguanidine and single photon emission computed tomography scan showed diffuse uptake of the radioisotope within the abdominal mass with no evidence of metastases. Figure 1b demonstrates the tumour as a multiplanar reformatting fusion image.

Paragangliomas, also referred to as extra-adrenal pheochromocytomas, are rare catecholamine-secreting tumours that may occur anywhere along the sympathetic paraganglionic chain [1]. The prevalence of phaeochromocytoma and paraganglioma is in the order of 0.2–0.6% [1].

The majority of paragangliomas are intra-abdominal, with the organ of Zuckerkandl being one of the most common sites [1]. Named for the Austrian anatomist Emil Zuckerkandl, the organ of Zuckerkandl, is a collection of chromaffin cells spanning from the inferior mesenteric artery to the bifurcation of the aorta [2].

Most paragangliomas are sporadic. Approximately 30–35% of paragangliomas may have an underlying germline mutation [3]. Current Endocrine Clinical Practice Guidelines recommend that all patients with paragangliomas undergo testing for succinate dehydrogenase (SDH) mutations [2]. However, routine genetic screening is costly and may not be readily available in many developing countries and resource-limited settings.

The patient received sequential alpha and beta-receptor blockade, with doxazosin and atenolol, respectively. Two weeks later, she underwent successful open surgical excision of the tumour. Unfortunately, genetic screening for SDH mutations is not readily available in South Africa and could not be offered to this patient. Close follow-up will occur with 3 monthly measurements of 24-hour urine metanephrine and normetanephrine for the first year followed by annual measurements thereafter to assess for disease recurrence.

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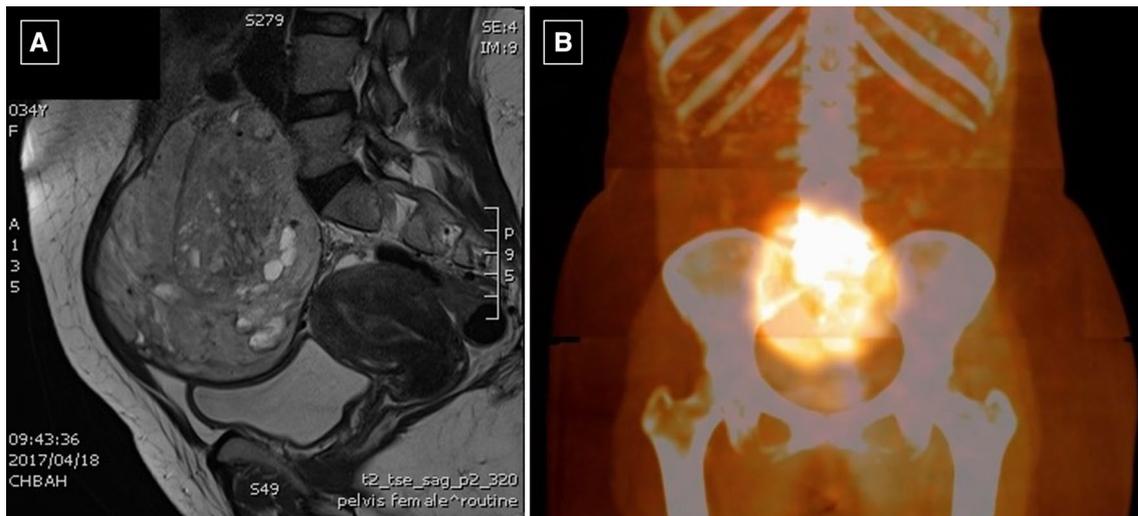


Fig. 1 **a** Sagittal MRI showing a large paraganglioma arising from the organ of Zuckerkandl. **b** Multiplanar reformatting fusion image demonstrates diffuse uptake of iodine-123 meta-iodobenzylguanine within the tumour

Compliance with ethical standards

Conflict of interest The author declares that they have no competing interests.

Statement of human and animal rights This article does not contain any studies with animal or human subjects performed by any of the authors.

Informed consent Obtained from the patient for their anonymized data to be published.

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