

# A glomus vagale tumour presenting with acute left ventricular failure

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## Abstract

A glomus vagale is a rare lesion of paraganglionic cells in the vagal perineurium. Secretion of hormones by such a tumour is exceedingly uncommon, although associated metabolic processes have been demonstrated. Clinical effects of secretion reported previously have been minimal. We report a case where resistant hypertension was eventually found to be due to a secretory glomus vagale, and was greatly alleviated by removal of the lesion.

**Key words:** Paranglioma; Neck; Hypertension

## Introduction

We report the case of a patient who presented with an episode of acute left ventricular failure caused by secretions from a glomus vagale tumour. Prior to the incident, she had suffered from long-standing intractable hypertension. Excision of the lesion led to a marked improvement of all cardiovascular problems. Glomus tumours are rarely secretory.<sup>1</sup> We believe this to be the first reported case where a glomus vagale tumour was revealed by cataclysmic vaso-active effects.

## Case report

A 51-year-old female was known to have suffered from hypertension for the previous three years. This had proved resistant to the standard intensive therapy of beta-blocker, angiotensin-converting enzyme inhibitor, diuretic and calcium antagonist.

While snorkelling in the sea off the Great Barrier Reef, she became acutely dyspnoeic shortly after entering the water. A severe tachycardia developed at the same time, and she began to cough frothy blood-stained sputum. These symptoms had resolved a little by the time she was admitted to hospital shortly afterwards, and continued to improve spontaneously. Acute left ventricular failure, involving pulmonary oedema, was diagnosed, with left ventricular regional wall abnormality on electrocardiography. The cause of this episode remained unidentified, and there were no further incidents of this kind, although recalcitrant hypertension persisted.

One year later, a swelling appeared at the angle of the mandible on the right, with medial displacement of the right tonsil. There were no cranial nerve defects. A computed tomography (CT) scan revealed a 50 mm mass in the right parapharyngeal space, and this was followed by magnetic resonance imaging (MRI) and digital subtraction angiography, which suggested a glomus vagale tumour supplied by the external carotid artery. Fine needle aspiration cytology, both per-orally and externally, was unhelpful. In view of these findings and the previous cardiovascular symptoms, plasma catecholamines were

measured, and a dopamine level of 3930 pmol/L was found, the normal being under 510 pmol/L. The urinary catecholamine level was 2370 pmol/L (normal upper limit 510 pmol/L). Serum adrenaline and noradrenaline levels were unremarkable. Screening for a second chemodectoma or pheochromocytoma proved negative, and venous sampling from the internal jugular showed no increase in catecholamines, presumably due to intermittent secretion.

Treatment options of surgery or radiotherapy were discussed, and it was decided to excise the lesion.

Phenoxybenzamine was administered for 10 days prior to surgery to provide alpha-blockade.

The lesion was approached via a medial mandibulotomy. Dissection was impaired by the intense vascular reaction around the tumour. Ligation of the external carotid artery and its branches was performed, and the internal carotid preserved following sharp dissection of the lesion from this artery. The vagus nerve disappeared into the tumour and was divided, and the hypoglossal, very closely related to the mass, was sacrificed at the skull base, then re-anastomosed.

The tumour was removed intact, measuring 55 mm by 35 mm. Histology confirmed a vagal paraganglioma, without malignancy.

A fresh sample of the tumour was sent for endocrine examination, showing high concentrations of noradrenaline (138.29 nmol/g, the upper limit of normal being 73.25 nmol/g) and dopamine (3.55 nmol/g, upper limit of normal 0.33 nmol/g). After surgery, dysphagia was troublesome, but resolved spontaneously over the following weeks. There was a markedly weakened voice due to right vocal fold palsy resulting from vagal nerve division. The hypoglossal anastomosis was unsuccessful. Urinary catecholamine levels rapidly reverted to normal at 195 pmol/L (upper limit 510 pmol/L), and hypertension became much more responsive to treatment. There have been no further cardiac problems.

Right thyroplasty was carried out to correct the voice six months later. Eighteen months after surgery, hypertension is well controlled by diuretic and beta-blocker.

## Discussion

A glomus vagale is a paraganglioma arising from chemoreceptor cell bodies associated with the vagus nerve.<sup>2</sup> Paragangliomata are tumours of neuro-ectodermal tissue outside the adrenal gland, and nests of paraganglionic cells within the perineurium of the vague nerve just below or at the ganglion nodosum are believed to be the site of origin of glomus vagale.<sup>3</sup> These lesions are rare, with only approximately 200 having been recorded since the original report in 1935.<sup>2</sup>

Secretory behaviour in paragangliomata is very infrequent, and has been reported just six times in vagal lesions prior to this example. In each case, noradrenaline was the catecholamine produced, of which dopamine is the immediate precursor. A secretory potential, if not activity, has however been identified in other paragangliomata, with positive tests for tyrosine hydroxylase, the pivotal enzyme in the production of dopamine. This appears to have progressed to full-blown production rarely, and the clinical significance has been minimal.<sup>1</sup>

One case of a secretory glomus jugulare involved flushing and palpitations as well as hypertension, with grossly elevated dopamine levels.<sup>4</sup> The one previously reported dopamine-secreting glomus vagale caused no symptoms or signs.<sup>1</sup>

Our patient was remarkable in her presentation. She had attended physicians for several years with an unusually recalcitrant form of hypertension, advancing rapidly through the usual levels of pharmacotherapy to little or no avail. Her cataclysmic event, an episode of left ventricular failure, which occurred acutely when swimming in the sea, had remained unexplained. However, the possibility of an exaggerated vagal response to immersion in cool water when snorkelling, especially when this involved breath-holding, has been suggested by the physicians.

She has had no problems with immersion since, remains well, and is in full-time employment. The voice is now reasonable, dysphagia is minimal, and the unilateral hypoglossal deficiency has not proved problematical.

- **This is a case report of a patient with long-term hypertension who was found to be suffering with a glomus vagale**
- **The tumour was hormone-secreting**
- **The hypertension was alleviated by surgical resection of the tumour**

## References

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