

## CT-guided biopsy of suspected malignancy: A potential pitfall

**S Henderson**,<sup>1</sup> MB ChB; **W Elsaify**,<sup>2</sup> MB ChB, MSc, FRCSI, FRCS (Glasgow), FRCS (General Surgery)

<sup>1</sup> Department of Radiology, Victoria Infirmary, Glasgow, UK

<sup>2</sup> Department of Endocrine Surgery, James Cook University Hospital, Middlesbrough, UK

*Corresponding author: S Henderson (steven.henderson.04@aberdeen.ac.uk)*

Paragangliomas are rare catecholamine-secreting neuro-endocrine tumours that can arise from sympathetic or parasympathetic tissue. Any manipulation of these tumours, without appropriate medical therapy, can result in excess catecholamine release leading to a catecholamine crisis. Neuro-endocrine tumours must be considered prior to interventional biopsy of an unknown soft-tissue mass, and appropriate biochemical investigations should be performed in suspected cases to prevent catastrophic complications.

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A 70-year-old woman was referred for computed tomography (CT)-guided biopsy of an incidentally detected partially necrotic 35 mm para-aortic mass in the left infrarenal area (Figs 1 and 2). Immediately post procedure, she developed severe central chest pain. Anterolateral myocardial infarction was confirmed on ECG and serum biochemistry, and the patient underwent percutaneous coronary intervention. The lesion was histologically identified as a paraganglioma. Subsequent 24-hour urine collection showed markedly raised urinary adrenaline and noradrenaline as well as raised plasma normetadrenaline. The patient subsequently underwent laparotomy under appropriate medical therapy and the lesion was successfully excised.

### Discussion Background

A phaeochromocytoma is a rare neuro-endocrine tumour of the paraganglionic chromaffin cells affecting 2 - 8 persons per million per year.<sup>[1-6]</sup> Although usually benign, around 10% are found to be malignant.<sup>[1,2,7]</sup> They can arise in the adrenal medulla and also, in around 10% of cases, in the extra-adrenal paraganglionic cells.<sup>[1,3-5]</sup> These extra-adrenal tumours are known as paragangliomas.

Paragangliomas can be classified as either sympathetic or parasympathetic, depending on the nature of the tissue from which they arise. Parasympathetic paragangliomas arise principally in the thorax, head and neck and, in around 95% of cases, are not hormone secretors.<sup>[4-6]</sup> Sympathetic paragangliomas, on the other hand, can arise from anywhere along the sympathetic nervous system from the skull base to the prostate gland. Most commonly, in around 75% of cases, they originate in the para-aortic region of the abdomen. They are frequently seen in the organ of Zuckerkandl, near the inferior mesenteric artery origin.<sup>[4]</sup> Typically, these sympathetic tumours secrete catecholamines.

It should be noted that phaeochromocytomas, arising from the adrenal medulla, are, by definition, sympathetic and therefore also always secrete catecholamines.<sup>[2,4,6-9]</sup>

### Clinical presentation and investigations

The majority of paragangliomas present in the third to fifth decades.<sup>[4]</sup> Paragangliomas can cause symptoms as a result of direct pressure<sup>[4]</sup> or because of hormone secretion. Tumours secreting catecholamines often present nonspecifically with headache, palpitations and often paroxysmal hypertension.<sup>[1,3,4,6,7,10]</sup> Notably, however, it is a rare cause of hypertension with less than 0.1% of hypertensives being found to have a secreting tumour.<sup>[1,10]</sup> In severe cases, arrhythmias, stroke, heart failure, pulmonary oedema, Raynaud's phenomenon and metabolic acidosis may be present.<sup>[2-4,7]</sup> In up to 40% of cases, however, these tumours are asymptomatic and discovered incidentally during the course of investigation for an unrelated condition.<sup>[2-4,6]</sup> If clinically suspected, diagnosis can be made with 24-hour urine collection demonstrating raised urinary catecholamines and metanephrines, which has a sensitivity of 87 - 90% and specificity >99%.<sup>[6]</sup> Serum catecholamines are also found to be raised on serum analysis although this is of less diagnostic benefit in modern practice owing to a high false positive rate.<sup>[1,3,4,7,10]</sup>

### Radiological investigation

Radiologically, CT, MRI, metaiodobenzylguanidine scan and PET imaging are all used in locating the primary lesion and the detection of metastatic disease.<sup>[4-6]</sup> On cross-sectional imaging, the lesions are usually visible although they usually appear as a non-specific soft-tissue mass.<sup>[10]</sup>

CT-guided biopsy is generally contra-indicated in suspected cases of paraganglioma. Such procedures can cause a surged release of catecholamines into the systemic circulation, which can lead to features of a catecholamine crisis – headache, sweating, elevated blood pressure,

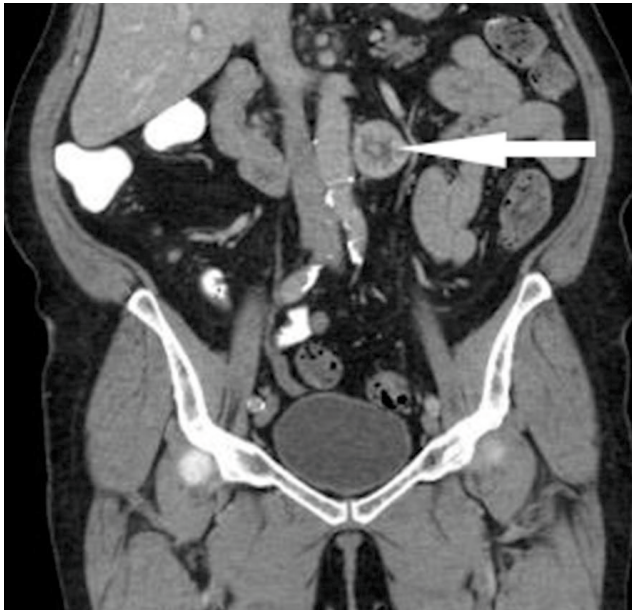


Fig. 1. Coronal CT slice through abdomen showing mass indicated by arrow.



Fig. 2. Axial CT slice through abdomen showing mass indicated by arrow.

lesion haemorrhage, haemodynamic compromise, and limb and cardiac ischaemia.<sup>[3,6,7,10]</sup> Cases of death precipitated by catecholamine release following CT-guided biopsy of adrenal pheochromocytomas have been reported.<sup>[3]</sup>

## Conclusion

Thirteen per cent of patients with a catecholamine-secreting tumour will not be hypertensive and 8% will be completely asymptomatic.<sup>[3]</sup> These figures, coupled with the nonspecific imaging features, make it imperative that, prior to performing biopsy, paraganglioma is considered and, if considered a realistic possibility, excluded biochemically. This can be done with 24-hour urine collection as described above. There should be a low threshold for screening any extra-adrenal mass planned for intervention. Certainly, all patients who undergo biopsy of an adrenal mass must be screened for pheochromocytoma.<sup>[10]</sup> In the case of a positive screening result, biopsy is unlikely to influence the diagnosis and would be actively contra-indicated.

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