CT-guided biopsy of suspected malignancy: A potential pitfall

S Henderson,[1] MB ChB; **W Elsaify**,[2] MB ChB, MSc, FRCSI, FRCS (Glasgow), FRCS (General Surgery)

[1] *Department of Radiology, Victoria Infirmary, Glasgow, UK*
[2] *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.

S Afr J Rad 2013;17(4):139-140. DOI:10.7196/SAJR.917

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.[1-6] Although usually benign, around 10% are found to be malignant.[1,2,7] They can arise in the adrenal medulla and also, in around 10% of cases, in the extra-adrenal paraganglionic cells.[1,3-5] 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.[4-6] 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.[4] 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.[2,4,6-9]

Clinical presentation and investigations

The majority of paragangliomas present in the third to fifth decades.[4] Paragangliomas can cause symptoms as a result of direct pressure[4] or because of hormone secretion. Tumours secreting catecholamines often present nonspecifically with headache, palpitations and often paroxysmal hypertension.[1,3,4,6,7,10] Notably, however, it is a rare cause of hypertension with less than 0.1% of hypertensives being found to have a secreting tumour.[1,10] In severe cases, arrhythmias, stroke, heart failure, pulmonary oedema, Raynaud's phenomenon and metabolic acidosis may be present.[2-4,7] In up to 40% of cases, however, these tumours are asymptomatic and discovered incidentally during the course of investigation for an unrelated condition.[2-4,6] 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%.[6] 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.[1,3,4,7,10]

Radiological investigation

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

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,

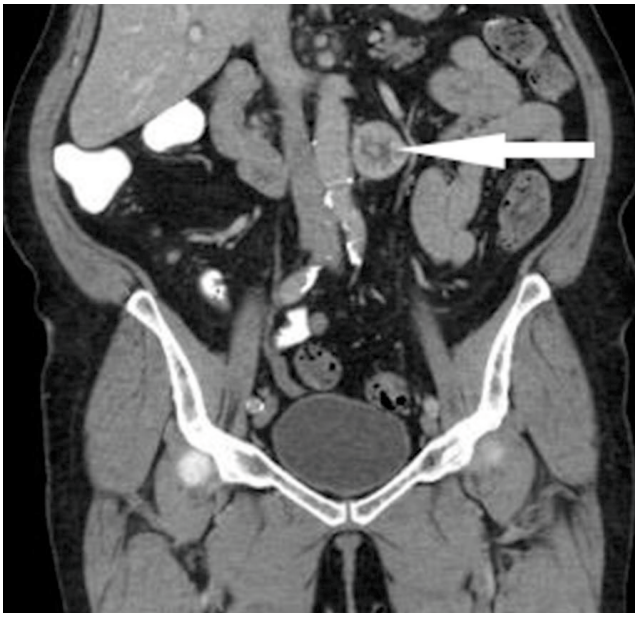

Fig. 1. Coronal CT slice through abdomen showing mass indicated by arrow.

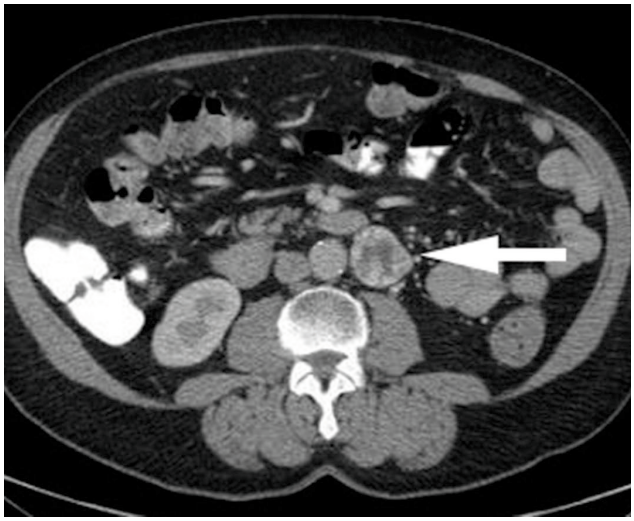

Fig. 2. Axial CT slice through abdomen showing mass indicated by arrow.

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

Conclusion

Thirteen per cent of patients with a catecholamine-secreting tumour will not be hypertensive and 8% will be completely asymptomatic.[3] 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 phaeochromocytoma.[10] In the case of a positive screening result, biopsy is unlikely to influence the diagnosis and would be actively contra-indicated.

1. Rangaswamy M, Kumar SP, Asha M, Manjunath GV. CT-guided fine needle aspiration cytology diagnosis of extra-adrenal phaeochromocytoma. J Cytol 2010; 27(1):26-28.
2. Welander J, Soderkvist P, Gimm O. Genetics and clinical characteristics of hereditary phaeochromocytomas and paragangliomas. Endocr Relat Cancer 2011;18:R253-R276. [http://dx.doi.org/10.1530/ERC-11-0170]
3. Sood SK, Balasubramanian SP, Harrison BJ. Percutaneous biopsy of adrenal and extra-adrenal retroperitoneal lesions: Beware of catecholamine secreting tumours! The Surgeon 2007;5(5):279-281.
4. Lee JA, Duh Q. Sporadic paraganglioma. World J Surg 2008;32:683-687. [http://dx.doi.org/10.1007/s00268-007-9360-4]
5. Havekes B, King K, Lai EW, et al. New imaging approaches to pheochromocytomas and paragangliomas. Clin Endocrinol 2010;72(2):137-145. [http://dx.doi.org/10.1111/j.1365-2265.2009.03648.x]
6. Gimm O, DeMicco C, Perren A, et al. Malignant pheochromocytomas and paragangliomas: A diagnostic challenge. Arch Surg 2012;397:155-177. [http://dx.doi.org/10.1007/s00423-011-0880-x]
7. Bajwa SJS, Bajwa SK. Implications and considerations during pheochromocytoma resection: A challenge to the anaesthesiologist. Indian J Endocrinol Metab 2011;15(Suppl4):S337-S344. [http://dx.doi.org/10.4103/2230-8210.86977]
8. Pacek K. Pheochromocytoma: A catecholamine and oxidative stress disorder. Endocr Regul 2011;45(2):65-90.
9. Levy MT, Braun JT, Pennant M, Thompson LDR. Primary paraganglioma of the parathyroid: A case report and clinopathologic review. Head Neck Pathol 2010;4(1):37-43. [http://dx.doi.org/10.1007/s12105-009-0157-7]
10. Paulsen SD, Nghiem HV, Korobkin M, Caoili EM, Higgins EJ. Changing role of imaging-guided percutaneous biopsy of adrenal masses: Evaluation of 50 adrenal biopsies. AJR 2004;182(4):1033-1037.