Rare disease

Management of adrenal incidentaloma: size still matters

Jordan ST Bowen,¹ Andrew Parker,² Shubha S Bellur,² Bangalore Anilkumar,³ David Dove,⁴ Greg P Sadler,⁵ Upendram Srinivas-Shankar⁶

¹Oxford Deanery, Heatherwood and Wexham Park Hospital, Slough, Berkshire, UK;

²Department of Pathology, John Radcliffe Hospital NHS Trust, Headington, Oxford, UK;

³Department of Radiology, Heatherwood and Wexham Park Hospital, Slough, UK;

⁴Department of Diabetes and Endocrinology, Heatherwood and Wexham Park Hospital, Slough, Berkshire, UK;

⁵Department of Surgery, John Radcliffe Hospital NHS Trust, Headington, UK;

⁶Department of Diabetes and Endocrinology, St Helens and Knowsley Teaching Hospitals NHS Trust, St Helens, UK

Correspondence to Dr Upendram Srinivas-Shankar, usrinivas@yahoo.com

Summary

A 56-year-old man was found to have an adrenal incidentaloma on a CT scan of the abdomen. Clinically and biochemically, the mass was not functional. MRI scan revealed a heterogeneously enhancing, T2-hyperintense, right-sided adrenal mass (4.5×6.5 cm). Meta-iodo-benzylguanidine scan was normal, making a diagnosis of pheochromocytoma unlikely. As the mass was larger than 4 cm, it was excised and histopathological examination revealed a rare, composite tumour: benign adrenal adenoma with haemangiomatic and myelolipomatous components. This case highlights the difficulties encountered by a clinician faced with investigating a potentially malignant adrenal mass (based on size) and correlates radiological findings with a rare histopathological specimen.

BACKGROUND

This case is interesting because it is, as far as we can tell, the first record of its histological tumour-type. We suspect that the prevalence is under-reported and will become more manifest with the increased detection of adrenal incidentalomas, with increasing use of radiological imaging of the abdomen. Further, it provides an opportunity to document

the radiological-pathological correlation, which may aid future radiological characterisation of adrenal masses.

CASE PRESENTATION

A 56-year-old man was referred to the endocrine outpatient clinic as he was found to have an adrenal mass on a CT scan performed for the evaluation of haematuria. He reported

Figure 1 Postcontrast, T1 weighted MRI image showing the right adrenal gland with peripheral and central nodular enhancement.

Figure 2 Macroscopic image of the solid cystic tumour with the compressed rim of the normal adrenal gland.

Figure 3 Tumour showing cortical adenoma and myelolipoma (H&E; magnification x4).

intermittent sweating, fatigue and unintentional weight loss. He denied palpitations, headache, dizziness, abdominal pain or diarrhoea. He was an ex-smoker and had hypertension. On examination, he appeared well, with a pulse rate of 80/min, and blood pressure of 130/70 mm Hg; without postural drop in blood pressure. He did not have neurofibromatosis or clinical features of Cushing's syndrome.

INVESTIGATIONS

Routine investigations including full blood count, serum calcium, renal, liver and thyroid function tests were normal. Serum renin and aldosterone ratio and 24 h urine free cortisol excretion were normal. Twenty four hours

urine collection revealed mildly raised total metadrenaline (4.64 $\mu\text{mol}/24\text{ h}$; normal range (NR) $<3.70\text{ }\mu\text{mol}/24\text{ h}$), normetadrenaline (3.60 $\mu\text{mol}/24\text{ h}$; NR $<2.50\text{ }\mu\text{mol}/24\text{ h}$) and norepinephrine (554 nmol/24 h, NR $<500\text{ nmol}/24\text{ h}$) on one occasion, however repeat collections were normal. Meta-iodobenzyl guanidine (MIBG) scan was normal making a diagnosis of pheochromocytoma very unlikely. MRI scan revealed a heterogeneously enhancing, predominantly T2-hyperintense, right adrenal mass (4.5 $\times$ 6.5 cm). There was high signal within the lesion on T1 sequence and minimal loss of signal, on out-of-phase imaging peripherally, with heterogeneous peripheral enhancement (figure 1).

Figure 4 Tumour showing areas of cavernous haemangioma and cortical adenoma (H&E x2).

Histopathological examination revealed an enlarged adrenal gland with a cortical tumour confined to the capsule, containing multiple cystic spaces with blood clots (figure 2). Microscopically the tumour comprised, lipid-rich, zona fasciculata-like cells dispersed in an organoid pattern, and cystic spaces with blood clots, representing thin-walled, endothelium-lined cavernous vascular spaces. Some vascular spaces were filled with blood lakes and others showed large thrombi in various stages of organisation and osseous metaplasia. The histological features were suggestive of a combined adrenal cortical adenoma with cavernous haemangioma as well as focal areas of mature adipose tissue with haematopoietic elements, indicating a myelolipomatous component. There were occasional bizarre nuclei but no mitotic figures and no vascular or capsular invasion (figures 3 and 4).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included benign (adrenal adenomas, lipomas) and malignant adrenal masses (adrenal carcinomas and metastasis). Further endocrine hormone secreting masses (Conn's adenoma, cortisol-producing masses and pheochromocytomas) need to be excluded.

TREATMENT

The patient underwent laparoscopic adrenalectomy on the right side.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful and the patient remained well.

DISCUSSION

The detection of adrenal incidentalomas (usually ≥ 1 cm in diameter) is increasing (0.3 to 4%) with widespread use of abdominal imaging.¹ The clinician needs to determine, if the mass is functional and whether it is benign or malignant. Biochemical investigation for hypercortisolism (Cushing's syndrome), hyperaldosteronemia and pheochromocytoma are necessary.² Over 40% of adrenal incidentalomas are cortical adenomas. Pheochromocytomas may comprise up to a tenth of cases.³ Adrenal carcinoma occurs in up to 5% and metastases in the adrenal gland (lung, kidney, gastrointestinal tract, breast and liver) occur in up to 2% of adrenal incidentalomas (usually bilateral). One per cent of adrenal adenomas produce aldosterone (Conn's syndrome). The remainder are myelolipomata and other rare tumour types.

MRI and CT features of the adrenal mass may aid in differentiation between benign and malignant adrenal adenomas. The lack of radiological and pathological correlation of rare composite adrenal tumours (like our case), contributes to the uncertainty in the characterisation of benign adrenal masses based on imaging alone.⁴ Nevertheless, a cut-off size of 4 cm has been accepted as a differentiator between benign and malignant masses: 6% of masses over 4 cm are malignant and the risk of malignancy rises to 25% in masses over 6 cm (sensitivity 93%, specificity 25% and positive predictive value 25%).⁵ In such cases excision of the adrenal mass is considered obligatory. Our case illustrates the first mention in literature of an adrenal adenoma with haemangioma and myelolipomatous components and aids in the correlation between radiological imaging and histological findings.

Learning points

- ▶ The detection of adrenal incidentalomata is increasing with widespread use of abdominal imaging.
- ▶ Composite tumours are a rare cause of adrenal incidentaloma, such as the adrenal adenoma and haemangioma with myelolipoma described here.
- ▶ Greater correlation between imaging and tumour-type may improve the characterisation of adrenal masses by radiological imaging (and reduce the need for surgical intervention).

Competing interests None.

Patient consent Obtained.

REFERENCES

1. **Bovio S**, Cataldi A, Reimondo G, *et al*. Prevalence of adrenal incidentaloma in a contemporary computerized tomography series. *J Endocrinol Invest* 2006;**29**:298–302.
2. **Grumbach MM**, Biller BM, Braunstein GD, *et al*. Management of the clinically inapparent adrenal mass ("incidentaloma"). *Ann Intern Med* 2003;**138**:424–9.
3. **Mansmann G**, Lau J, Balk E, *et al*. The clinically inapparent adrenal mass: update in diagnosis and management. *Endocr Rev* 2004;**25**:309–40.
4. **Otal P**, Escourrou G, Mazerolles C, *et al*. Imaging features of uncommon adrenal masses with histopathologic correlation. *Radiographics* 1999;**19**:569–81.
5. **Alexandraki KI**, Grossman AB. Adrenal incidentalomas: 'the rule of four'. *Clin Med* 2008;**8**:201–4.

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