Rare disease

Diaphoresis and abdominal pain caused by extra-adrenal paragangliomas

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Summary

A 63-year-old lady presented with suprapelvic pain, weight loss and night sweats. On examination, she was noted to be hypertensive with a distended abdomen. Imaging (CT) revealed a 9.5 cm retroperitoneal mass with a high degree of vascularity and necrotic centre. The patient's urinary and plasma catecholamines were significantly raised and subsequent radio-isotope scan suggested the tumour was likely to be of a neuroendocrine nature. A diagnosis of a malignant paraganglioma was made. Malignant paragangliomas derive from sympathetic tissue and secrete catecholamines. Diagnostic uncertainty might lead to biopsy of tumour but this carries a high-risk of catecholamine-induced complications such as hypertensive crisis, cardiac arrhythmias and cardiac ischaemia and must be avoided.

BACKGROUND

1. Paragangliomas are an important differential for retroperitoneal tumour.
2. Radiologically, paraganglionomas may be mistaken for lymphoma, however, percutaneous biopsy is contraindicated due to risk of life-threatening haemorrhage or hypertensive crisis.
3. The different types of catecholamines secreted by paragangliomas is a complicated field and revisiting the subject is helpful.

CASE PRESENTATION

A 63-year-old woman presented to the emergency department with an 18-month history of weight loss, night sweats and dull suprapubic pain radiating to her back. This pain was present throughout the day, was constant and dull in nature, but not positional or related to food. Examination demonstrated a non-tender, distended abdomen, but the rest of the examination was unremarkable. Blood pressure was 142/65 mm Hg and pulse 95/min. Bloods, including haemoglobin and inflammatory markers were in normal range.

The history was sufficiently prolonged and atypical, that together with a grossly abnormal examination finding of a distended abdomen, an axial CT imaging study was requested.

INVESTIGATIONS

Axial CT imaging demonstrated a 9.5 cm retroperitoneal mass that was highly vascular with a necrotic centre (figure 1) and these features made it highly suspicious for a neuroendocrine tumour. The patient was scheduled for CT guided biopsy to exclude lymphoma but after senior radiographic review, measurement was made of catecholamine secretion. Urinary (36.1 mol/dl; reference <3.30) and plasma (14.49 nmol/l; reference <1.3) normetanephrine were elevated but urinary (0.38 mol/dl; reference <1.2) and plasma (0.12 nmol/l; reference <0.7) metanephrine were within the normal range. A meta-iodobenzylguanidine scan confirmed high uptake in the retroperitoneal mass

plus further uptake in the region of the seventh thoracic vertebra (figure 2), confirming the suspected diagnosis of an extra-adrenal paraganglioma.

DIFFERENTIAL DIAGNOSIS

The differentials for this axial CT image include lymphoma, gastrointestinal tumour or neuro-endocrine tumour.

TREATMENT

After pharmacological α and β blockade, the patient underwent oncological resection.

Histopathology confirmed a malignant paraganglioma. Urinary and plasma normetanephrine levels normalised postoperatively.

DISCUSSION

Paragangliomas (PGLs) derive from either sympathetic tissue (sPGL) in adrenal and extra-adrenal locations, or from parasympathetic tissue (pPGL) of the head and neck.

Adrenal paraganglionomas are referred to as pheochromocytomas.

Figure 1 Contrast axial CT abdomen showing a large, vascular, retroperitoneal mass with a necrotic centre (arrow).

Figure 2 Four hour whole-body image showing I123-mIBG avid large abdominal mass with a central area of heterogeneity. There is further uptake in the mid-thoracic region centrally (arrows).

Biochemically, extra-adrenal sPGL may often be distinguished from pheochromocytomas due to the preferential secretion of norepinephrine over epinephrine.

Extra-adrenal paraganglionomas are deficient in the enzyme PNMT (phenylethanolamine N-methyltransferase), which converts norepinephrine to epinephrine, whereas PNMT is found within the adrenal medulla. However, exclusive increases in plasma free normetanephrine can-

not be relied upon to indicate an adrenal or extra-adrenal location of a sPGL.

The classic triad of headache, sweating and palpitations occurs less frequently in sPGLs than is generally assumed (no more than 25%¹). The variable symptomatology may reflect the type of catecholamines secreted: norepinephrine acts primarily on α -adrenergic receptors and so patients with predominant norepinephrine secretion (as with this case) may not present with β -mediated symptoms (headaches, palpitations and anxiety) but with symptoms more characteristic of α -adrenergic activity such as diaphoresis and/or hypertension. Due to the absence of typical symptoms of catecholamine excess, the diagnosis of paraganglionoma is often delayed until an advanced stage of the disease and patients may then only present with symptoms caused by a tumour mass effect.

Diagnostic uncertainty may also lead to tumour biopsy but this carries a high-risk of catecholamine-induced complications such as hypertensive crisis, cardiac arrhythmias and cardiac ischemia and must be avoided.

This case highlights the fact that paragangliomas are an important differential for a retroperitoneal tumour. Radiologically, paragangliomas may be mistaken for lymphoma, however, percutaneous biopsy is contraindicated due to risk of life-threatening haemorrhage or hypertensive crisis.

Learning points

- Paragangliomas are an important differential for retroperitoneal tumour.
- Radiologically, paragangliomas may be mistaken for lymphoma, however, percutaneous biopsy is contraindicated due to risk of life-threatening haemorrhage or hypertensive crisis.

Competing interests None.

Patient consent Obtained.

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