obstruction. Multiple strictures (2–25) are often present, with frequent relapse following resection. Steroid responsiveness is typical but steroid dependence often occurs. Limited experience with immunosuppressants (azathioprine and methotrexate) suggests lack of benefit.⁴ Weight loss was a feature in about half of the cases described and systemic symptoms such as fever and arthralgia may also be present. Iron deficiency anaemia has not, we think, been previously described as the presenting complaint.

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A tense case— Carney's triad

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A rare differential diagnosis for secondary hypertension is Carney's triad—extra-adrenal paraganglioma, gastric leiomyosarcoma, pulmonary chondroma.¹

CASE HISTORY

A man aged 46 with known refractory hypertension was referred for investigation after a year of occipital headaches on exertion and six months of increasing lethargy and fatigue. Associated symptoms included drenching night sweats, paraesthesiae in his fingers, cold peripheries, a decreasing libido, and weight loss of 7 kg over the past four months. Current treatment was felodipine 10 mg and lisinopril 10 mg daily.

Blood pressure on admission was 162/102 mmHg; routine blood indices were all normal apart from mildly

deranged liver function tests. 24-hour urinary excretion of catecholamines and their metabolites was measured by high-performance liquid chromatography with solid phase extraction while the patient was on the original doses of felodipine and lisinopril. Norepinephrine was 20 000 nmol/24 h (normal range 0–500), epinephrine 24 nmol/24 h (0–100), normetanephrine 25.2 μ mol/24 h (0.1–3.3), dopamine 5480 nmol/24 h (0–3000) and vanillylmandelic acid 114 μ mol/24 h (<35). Chest radiography revealed a 5 cm calcific lesion in the left upper zone consistent with a pulmonary chondroma (Figure 1). This pulmonary lesion was not a new finding. It had been detected at age 26 when the patient had a routine chest radiograph after a road traffic accident in Italy; at that time he had declined further investigation. Ultrasound of the abdomen showed an enlarged fatty liver; the left kidney was mildly hydro-nephrotic but no masses were seen in the upper pole of either kidney. CT revealed a left-sided 7.5 cm para-aortic mass consistent with an extra-adrenal paraganglioma (Figure 2). An iodine-123-meta-iodobenzylguanidine (MIBG) scan was negative, possibly because labetalol had been prescribed earlier and intravenous phenoxybenzamine had been given in preparation for surgery. An attempt was then made at resecting the tumour. Frozen and paraffin sections confirmed that it was a pheochromocytoma with lymph node metastasis. Unfortunately, postoperative CT revealed residual tumour not amenable to further surgery. Endoscopy was performed to exclude gastric leiomyosarcoma. No such lesion was seen and biopsies were negative. Postoperatively, the urinary catecholamine excretion did not become normal but the patient is at present symptom-free on a regimen of sandostatin 50 μ g and phenoxybenzamine 10 mg twice daily and propranolol 40 mg, lisinopril 20 mg and bendrofluazide 2.5 mg daily.

Figure 1 Chest radiograph showing pulmonary chondroma in left upper zone

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COMMENT

The diagnosis of Carney's triad, which was characterized in 1977, requires at least two of three components—gastric leiomyosarcoma, extra-adrenal paraganglioma and pulmonary chondroma¹. In the latest review, by Carney in 1999, cases with two or three of the elements had been reported in 79 patients, 84% of them female.² The neoplasms can appear at any age and in any sequence. 78% of patients had two tumours on presentation, the most frequent combination being gastric leiomyosarcoma and pulmonary chon-

droma.² To date, only two patients have shown a complete Carney's triad at first presentation.³ Typical early features are chronic iron deficiency anaemia or gastrointestinal bleeding.⁴ Only 4% of patients are detected through hypertension secondary to a catecholamine-secreting extra-adrenal paraganglioma.² Carney's triad needs to be thought of in patients with such tumours. The pulmonary chondromas have high calcium content and are often picked up incidentally on chest radiography. These are benign and resection is unnecessary.⁵ Gastric leiomyosarcomas, by contrast, are potentially fatal. Gastroscopy with biopsy is the investigation of choice. Early operation is desirable since this tumour is insensitive to chemotherapy and radiotherapy.⁶ A functioning extra-adrenal paraganglioma can be picked up by measurement of urinary catecholamines and the tumour is often delineated by CT, ultrasonography or MRI. Additional information can be had from iodine-123-meta-iodobenzylguanidine scanning (preferably not performed after treatment with intravenous phenoxybenzamine or beta blockade) or from indium-111-octreotide scanning.⁵ Again, surgery is the best treatment, to prevent life-threatening dysrhythmias, hypertensive crises and severe end-organ damage.⁴

In the patient reported here, the detection of a pulmonary chondroma at age 26 should ideally have triggered further investigations at the time. The paraganglioma might then have been more amenable to surgical intervention. He will now require lifelong treatment together with follow-up to monitor the status of his paraganglioma and exclude the development of gastric leiomyosarcoma.

Figure 2 CT showing an irregularly shaped left para-aortic mass with central ring calcification (a) and slight enhancement following contrast (b)

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