Secretory symptoms from metastatic adrenal cortical carcinoma responding to octreotide

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Summary
The prognosis of patients with metastatic adrenal cortical carcinoma is poor, and their disabling symptoms are usually unresponsive to conventional therapy. A patient with Cushing’s syndrome secondary to a secretory adrenal cortical carcinoma was treated with octreotide, endocrine therapy and chemotherapy having failed. Treatment led to a dramatic relief of her symptoms with a fall in corticosteroid secretion. Somatostatin analogue therapy for this tumour should be encouraged in view of the lack of alternative palliative treatment.

Keywords: octreotide, adrenal cortex, carcinoma

Introduction
Adrenal cortical carcinoma is a rare cause of Cushing’s syndrome. At least half of all patients have metastatic disease at presentation. No adjuvant treatment is effective, and relapse is followed by a median survival of approximately three months. There is no effective cytotoxic drug or drug combination available for recurrent adrenal cortical carcinoma; 30–40% of patients will have a partial response to treatment, but the median survival is four months. The management of locally recurrent or metastatic tumour is therefore aimed at palliating symptoms secondary to hormonal hypersecretion, visceral pain, or bone pain. Agents such as mitotane and aminoglutethimide given to reduce hypercortisolaeemia are relatively ineffective and are themselves toxic. There is a need for new treatments of this condition that are not toxic and which will palliate hypersecretory symptoms. For this reason we have explored the value of octreotide in adrenal cortical carcinoma.

Case report
A 46-year-old woman presented in May 1993 with weight gain, amenorrhoea, proximal myopathy, and severe acne. She had many of the clinical stigmata of Cushing’s syndrome and a right-sided abdominal mass. Her initial 24-hour urinary free cortisol levels (UFCs) were greater than 3000 (normal range 0–270 nmol/24 h) and her morning plasma cortisol level was over 800 (normal range 200–700 nmol/l) with loss of diurnal variation, but without suppres-

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Comment
Octreotide is effective at controlling endocrine hypersecretion and hormone-induced symptoms associated with a variety of functional endocrine tumours. In view of the lack of therapeutic options for adrenal cortical carcinoma and the broad spectrum of inhibitory endocrine effects of somatostatin analogues, octreotide was successfully applied as treat-
To inhibit aldosterone secretion. A marked decrease of plasma cortisol levels with infusional somatostatin was observed by Aynsley-Green et al in an evaluation of the influence of this peptide on insulin secretion. Octreotide has also been shown to reduce UFCs in patients with ACTH-dependent Cushing's syndrome, but the primary site of action of somatostatin could not be inferred from any of these studies.

In our patient, despite the negative Octreoscan, there was a rapid symptomatic and biochemical improvement in response to octreotide. The mechanism for this is unknown. The response to octreotide in ACTH-independent adrenal cell carcinoma demonstrated by the serial measurement of UFCs has not previously been reported. In view of the limited efficacy of current agents in adrenal cortical carcinoma, a trial of octreotide to control secretory symptoms in patients with this condition would seem reasonable.

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