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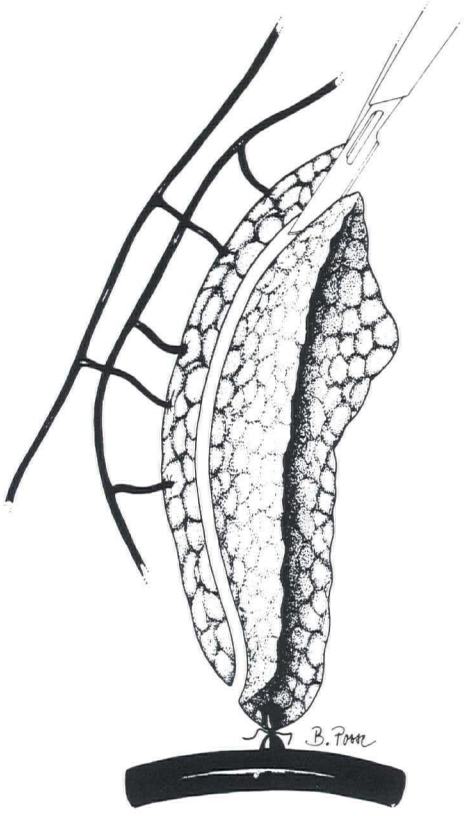
We report two patients in whom pheochromocytoma was treated by subtotal adrenalectomy leaving a rim of vascularized cortical tissue in situ. Both patients are doing well without cortisol supplementation although they have subnormal cortisol responses to ACTH stimulation. (Henry Ford Hosp Med J 1987;35:127-8)

The clinical management of adrenal disease in multiple endocrine neoplasia type 2 (MEN-2) is controversial. Bilateral total adrenalectomy has been suggested to prevent risk of recurrence. However, patients then require lifelong steroid medication. Although only minimal morbidity from bilateral adrenalectomy has been reported (1), this has not been our experience. An alternative is to leave the apparently unaffected adrenal gland in place. In a series of patients undergoing only unilateral adrenalectomy, it has been demonstrated that contralateral adrenalectomy could be postponed for several years (2). Total adrenalectomy and transplantation of cortical tissue has so far been successful only in patients with Cushing's syndrome. We report here our experiences with two patients with the MEN-2 syndrome on whom we performed subtotal adrenalectomy with the aim of removing all the adrenal medullary tissue.

Technique

The adrenal gland was widely exposed by a flank or dorsal approach. Part of the gland was mobilized and the adrenal vein ligated and divided. A rim of one adrenal gland, which by digital palpation was felt to be thin, soft, and nontumorous and into which several small vessels can be seen entering, was identified. The rest of the gland was resected leaving only this rim of vascularized cortical tissue in situ (Figure).



Figure—Schematic drawing of resection of the adrenal gland. A thin, well-vascularized rim of the adrenal gland is identified and left in situ and the rest of the gland is resected.

Case Reports

Case 1

A 31-year-old man had undergone a left adrenalectomy for pheochromocytoma nine years previously. At a follow-up examination his remaining adrenal gland was enlarged and hypersecreting. This right adrenal gland was resected with about one third of cortical tissue being left in situ. The specimen contained two pheochromocytomas, 2.5 and 1 cm in diameter. His urinary norepinephrine levels were within normal limits both preoperatively and postoperatively, but the epinephrine

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Table
Urinary Excretion of Norepinephrine, Epinephrine, and Cortisol after Subtotal Adrenalectomy

	Norepinephrine*		Epinephrine†		Cortisol‡
Patient	Preoperative	Postoperative	Preoperative	Postoperative	Postoperative
1	384	183	59	10	120
2	699	250	108	6	195

*Normal level < 400 mmol/24 hours.
†Normal level < 80 mmol/24 hours.
‡Normal level 80-300 mmol/24 hours.

concentration was almost nondetectable postoperatively (Table). His postoperative cortisol medication was withdrawn after one month. Since then, he has been without cortisol supplementation except for a few periods with fever. Two years postoperatively his urinary cortisol level is normal, but the response to ACTH stimulation is subnormal.

Case 2

A 31-year-old woman, who had been operated on for medullary thyroid carcinoma ten years earlier, was found to have a pheochromocytoma in the right adrenal gland. The tumor, which measured 3.5 cm in diameter, was removed along with the entire right adrenal gland. The left adrenal was resected leaving a rim of cortical tissue in situ. Urinary catecholamines were normal after operation (Table). Three months postoperatively cortisol medication was withdrawn and after another six months basal plasma cortisol and urinary cortisol levels were normal. However, her response to ACTH stimulation was subnormal.

Conclusions

In two patients with MEN-2 a rim of adrenal cortical tissue was left in situ when adrenalectomy was performed. Postoperatively, normal norepinephrine and very low epinephrine levels were detected in the urine. Both patients are doing well without cortisol supplementation. Two more patients have recently been operated on with good immediate clinical results. However, follow-up evaluations are not yet complete. In our opinion, subtotal adrenalectomy should be considered a valuable alternative in the treatment of patients with MEN-2.

Acknowledgments

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"Man is a prisoner of his ideas . . . if he does not possess suitable and necessary tools."

Claude Bernard—1865