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Surgical Approach of Synchronous Medullary Thyroid Carcinoma and Pheochromocytoma in MEN 2 Syndrome

Georg F. W. Scheumann,* and Henning Dralle*

In cases with concurrent medullary thyroid carcinoma (MTC) and pheochromocytoma, discussion regarding a one-stage versus two-stage treatment strategy approach remains open. From 1975 to 1990, 11 of 25 multiple endocrine neoplasia type 2 (MEN 2) patients presented with biendocrinopathies or triendocrinopathies synchronously. All patients were treated surgically and followed subsequently in our hospital. Of the group of nine patients with concurrent MTC and pheochromocytoma, five were treated in one-stage and four in two-stage procedures. No patient had major complications intraoperatively. For the two-stage group, the total hospital stay (preoperatively and postoperatively) averaged 35 days. For the one-stage group, the total hospital stay averaged 25 days. In patients with increased operative risks (patients with higher age and impaired physical condition or if neck surgery includes transsternal cervicomediastinal lymphadenectomy), two-stage procedures should be selected. However, in young patients with the MEN 2 syndrome or syndromes with small tumors detected by family screening, thyroidectomy, cervical lymphadenectomy, and adrenalectomy may be performed in a one-stage procedure without increasing surgically related morbidity. (Henry Ford Hosp Med J 1992;40:278-80)

The multiple endocrine neoplasia type 2 (MEN 2) syndrome appears in two variants. MEN 2A is defined as the MEN 2 syndrome without the mucosal neuroma phenotype, and MEN 2B as the MEN 2 syndrome with the mucosal neuroma phenotype (1). The majority of hereditary cases belong to the MEN 2A (70%) or familial non-MEN 2A variant (17% to 24%); only 6% to 13% are of the mucosal neuroma phenotype (MEN 2B) (2-4).

The syndrome is a genetically transmitted disease which predominantly affects the thyroid, parathyroid, and adrenal glands. Although surgery is generally regarded as the treatment of choice for each stage of the disease, the extent of surgical resection of the involved endocrine organs and the surgical time strategy in cases of synchronously involved glands is still debatable. In cases with concurrent medullary thyroid carcinoma (MTC) and hyperparathyroidism (HPT), the procedure with total thyroidectomy and parathyroidectomy performed in a one-stage operation is generally accepted because the extent of resection and operative risk for the patient does not differ from that for one-gland disease (2,5).

However, in cases with concurrent MTC and pheochromocytoma, discussion regarding treatment strategy (one-stage versus two-stage approach) remains open. We present our surgical approach in MEN 2 patients with regard to the synchronous appearance of MTC and pheochromocytoma.

Patients and Methods

From 1975 to 1990, 11 of 25 MEN 2 patients presented with biendocrinopathies or triendocrinopathies synchronously. All

patients were treated surgically and were followed subsequently at our hospital. The patients' data were stored, maintained, and evaluated by the Clinical Cancer Registry of the Medical School of Hannover. The information system used was developed and established under the DSM-11 (Digital Standard MUMPS) data base management and operating system. ("MUMPS" is the acronym for the Massachusetts General Hospital Utility Multi-programming System which was originally developed at the Laboratory of Computer Science at Massachusetts General Hospital.)

Results

In the 11 patients (44% of all MEN 2 patients) with synchronous biendocrinopathies or triendocrinopathies, two patients suffered from synchronous MTC and HPT (patients 202 and 304). Another two patients showed MTC, HPT, and pheochromocytoma (patients 101 and 503) at the time of clinical presentation. In seven patients, MTC and pheochromocytoma were demonstrated concurrently (Table). Four had bilateral pheochromocytoma. Bilateral pheochromocytoma occurred more frequently in patients with synchronous pheochromocytoma

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Table
Surgical Procedure in Nine Patients With Synchronous MEN 2/MTC and Pheochromocytoma (1975-1990)

Number	One-Stage	Day of Discharge	Complications	Number	Two-Stage	Day of Discharge	Complications
102	TT, parathyroidectomy, transperitoneal bilateral total adrenalectomy	17	—	101	December 1990, right translumbar total adrenalectomy	8	Intermittent bradyarrhythmia
					March 1991, TT	4	—
302	TT, parathyroidectomy, transperitoneal left total and right subtotal adrenalectomy	19	—	502	December 1986, TT	2	—
					July 1987, translumbar right total adrenalectomy	7	—
305	TT, parathyroidectomy, transperitoneal left and right subtotal adrenalectomy	24	—	503	March 1987, TT	8	—
					May 1988, transperitoneal bilateral total adrenalectomy with adrenocortical autotransplantation left forearm	15	—
601	TT, transperitoneal left total adrenalectomy	9	—	504	December 1987, transperitoneal right total adrenalectomy	22	Pneumonia
					April 1988, TT	4	—
1101	TT, transperitoneal left total adrenalectomy	13	—				

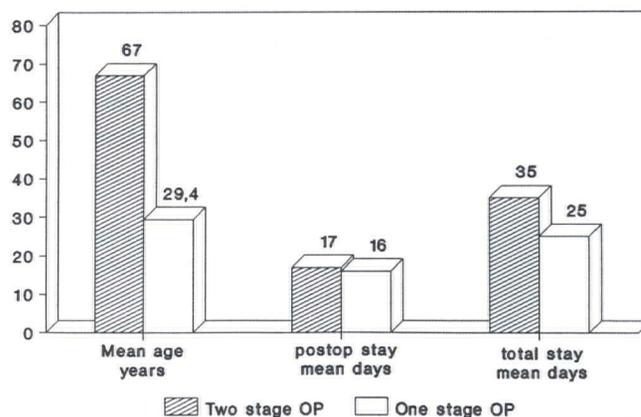
TT = total thyroidectomy.

and MTC, as opposed to those with metachronal pheochromocytoma and MTC (6).

Of the group of nine patients with concurrent MTC and pheochromocytoma, five were treated in a one-stage and four in a two-stage procedure. No patient had major complications intraoperatively. Preoperatively, all patients with synchronous MTC and pheochromocytoma were treated with a high dosage of α -receptor blockage (mean 270 mg phenoxybenzamine) for 13 to 25 days.

Two-stage group

The three females and two males had a mean age of 67 years (range 48 to 69 years). Two patients underwent thyroidectomy first because of symptomatic MTC (patients 502 and 503). They had been treated preoperatively with high dosage of α -receptor blockage without any intraoperative or postoperative complications. The two other patients were treated first by adrenalectomy and then by thyroidectomy. One patient had pneumonia after the primary transperitoneal unilateral adrenalectomy (patient 504) and was discharged from hospital on the 22nd postoperative day. Another patient experienced a short period of intermittent bradyarrhythmia immediately after unilateral translumbar adrenalectomy but without any delay in his hospital stay. The duration of postoperative care averaged 12 days for the single operative procedure and a total of 17.5 days for the two operative procedures for a total hospital stay (pre- and postoperative) averaging 35 days (Figure).



Figure—Comparison of patients' ages and length of hospital stays for one- and two-stage procedures.

One-stage group

The three females and one male had a mean age of 29.4 years (range 15 to 54 years). The surgical procedure in one-stage operations was cervical surgery followed by abdominal or lateral approach to the adrenal gland. There were no postoperative complications. The mean postoperative stay was 16.4 days with a total hospital stay (preoperatively and postoperatively) averaging 25 days (Figure).

Discussion

Surgical management of MEN 2 patients includes preoperative diagnostic procedures identifying the organs involved (7,8), preoperative medical treatment of patients with pheochromocytoma before adrenalectomy (9), and perioperative α -receptor blockade at the time of cervical intervention in patients with a pheochromocytoma diathesis not yet considered for adrenalectomy (6). Surgical strategy includes the management of patients with synchronous or metachronous multiorgan manifestations and the surgical technique of thyroid, parathyroid, and adrenal gland resection.

Of our MEN 2 patients, 44% presented with synchronous bi-endocrinopathies or triendocrinopathies. This group is heterogeneous, with an age range from 15 to 69 years, an MTC tumor stage variation from T1 to T4, and pheochromocytoma with or without clinical symptoms and unilateral or bilateral disease. Therefore, a general strategy for surgical procedure could not be given. The decision between one-stage or two-stage procedure with adrenalectomy first as routine procedure, or thyroidectomy first in the case of dominating malignant cervical disease, must be made individually based on the patient's physical and MEN 2-related condition if appropriate α -receptor blocking pretreatment is followed (9-11). In patients with increased operative risks (i.e., higher age, impaired physical condition, or if neck surgery includes transsternal cervicomediastinal lymphadenectomy), two-stage procedures should be selected. However, in young patients with the MEN 2 syndrome (12) and syndromes such as pheochromocytoma and small MTC detected by family screening, thyroidectomy, cervical lymphadenectomy, and adrenalectomy may be performed in a one-stage procedure with diminished individual discomfort attended with a second surgi-

cal intervention and without increased surgically related morbidity.

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