

The postoperative clinical course of pheochromocytoma in patients with multiple endocrine neoplasia type2 (MEN2)

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Introduction

Proper management of hereditary pheochromocytoma (Pheo) is a challenging issue because the disease often happens in bilateral adrenal glands. Recent clinical practice guidelines recommend partial adrenalectomy (PA) for bilateral and hereditary Pheo. The purpose of this study was to describe the postoperative prognosis of Pheo patients with MEN2.

Materials and methods

Patients : 59 Pheo patients with MEN2 underwent initial surgery in our hospital from 1982 to 2023.
Study design : Retrospective chart review of single institute
Research factor : Tumor location, size, surgical management, pathological findings, and prognosis

Outcome definition :

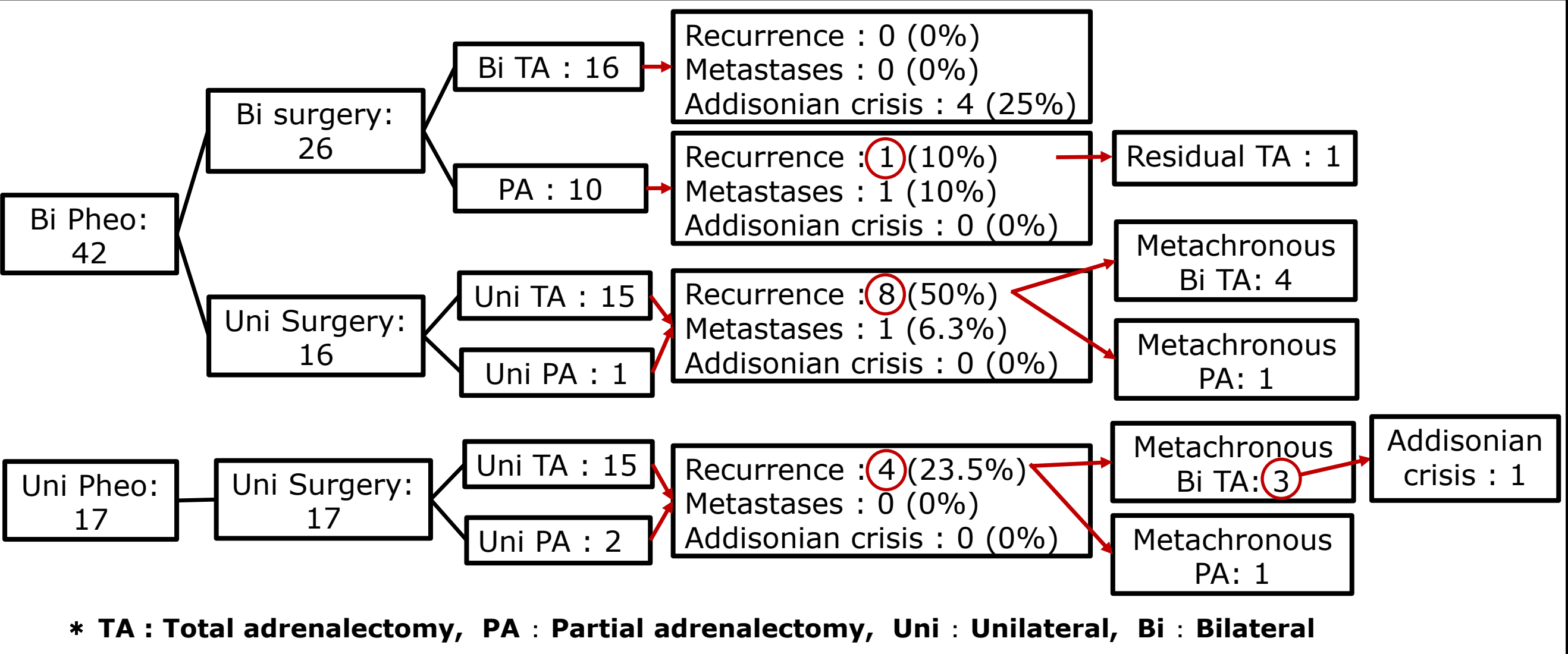
- Recurrence :** Residual adrenal recurrence
Sum of spot urine metanephrine (MN) and normetanephrine (NMN) ≥ 1.0 pg/mg·Cr + Tumor present
- Addisonian crisis :** Intravenous steroid administration was required
- Cortical function preservation :** Does not required postoperative corticosteroid replacement

Results

Patients Characteristics	
N	59
Female : Male	33 : 26
Age Median (Range)	34 (16 - 73) years
MEN type 2A : 2B	56 : 3
Tumor size Median (Range)	4.5 (1.2 - 14.0) cm
Observation period Median (Range)	113 (0 - 427) months

At initial operation ...

Bilateral Pheo : 42Pts
Unilateral Pheo : 17Pts



In Bilateral Surgery → Synchronous or Metachronous Bi TA : 24
Synchronous or Metachronous PA : 12

	TA (N = 24)	PA (N = 12)
Cortical function preservation	0 (0 %)	10 (83.3 %)
Recurrence	0 (0 %)	1 (8.3 %)
Addisonian crisis	5 (20.8 %)	0 (0 %)

3 of these patients had a rapid ACTH stimulation test within one week after surgery.

	Rapid ACTH stimulation test Peak cortisol	Cortical function preservation	Volume of preserved adrenal gland	Adrenal vein
Case 1	20.4 µg/dL	○	Unknown	Preserve
Case 2	3.6 µg/dL	×	25 %	Cut
Case 3	1.7 µg/dL	×	15 %	Cut

Discussion

The preservation rate of adrenocortical function for PA was high (83.3%), and the recurrence rate was low (8.3%). Furthermore, no Addisonian crisis developed during postoperative follow-up.

Bilateral TA carries little risk of recurrence, however, lifelong oral corticosteroids are mandatory, with a 20% frequency of Addisonian crisis.

Conclusion

The PA is appropriate as an alternative surgery to bilateral TA in selected patients with pheochromocytoma with MEN2.

What tests should be used to evaluate the preservation of adrenocortical function is a question for the future.