PP-A-17

THREE CASES OF CATECHOLAMINE-SECRETING NEUROENDOCRINE TUMOR IN A TERTIARY HOSPITAL, WEST JAVA, INDONESIA

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CASE
Pheochromocytomas or paragangliomas (PPGL) are rare neuroendocrine chromaffin-derived tumors. The annual incidence of PPGL is approximately 0.66 per 100,000 person-years. Approximately 50% of patients present with paroxysmal hypertension, 15-25% have the classic triad, while 5–15% are asymptomatic. Patients with a high degree of catecholamine excess may present with PPGL crisis with target organ complications.

Three cases of catecholamine-secreting neuroendocrine tumors (NET) were entertained in our institution. Patient I presented with a nonclassical triad, and patient II presented with a classic triad. Patient III was diagnosed as having a pheochromocytoma crisis, manifesting as recurrent myocarditis and cardiogenic shock. The patients were diagnosed as PPGL based on elevated metanephrine, adrenal CT scan, and Iodine-131-metaiodobenzylguanidine scintigraphy (I131-MIBG) positivity. Two patients underwent tumor resection after being perioperatively stable for 14 days, and one patient was lost to follow-up. The final diagnosis for patient I was progressive malignant paraganglioma, Patient II had pheochromocytoma. The patients were followed up and showed no residual symptoms after 1 month.

KEYWORDS
pheochromocytoma, paraganglioma, perioperative, adrenal-ectomy

PP-A-18

MULTIPLE ENDOCRINE NEOPLASIA TYPE 2A WITH ECTOPIC ACTH SYNDROME DUE TO BILATERAL PHEOCHROMOCYTOMA

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CASE
Ectopic ACTH syndrome (EAS) accounts for less than 5% of endogenous Cushing's syndrome. In a previous retrospective study on patients with endogenous Cushing's syndrome, only one had EAS from pheochromocytoma.

A 33-year-old Thai female with a cushingoid appearance complained of abdominal pain. CT scan showed an enlarged bilateral adrenal gland. Urine VMA was positive. 1 mg dexamethasone suppression test and 24 hr urine-free cortisol suggested endogenous hypercortisolism. Plasma ACTH was 21 pg/ml, not suppressible on high-dose dexamethasone suppression test. She was subsequently diagnosed with bilateral pheochromocytoma with ectopic ACTH and underwent bilateral adrenalectomy.

Her genetic testing showed a heterozygous pathogenic variant in the RET gene (c.1901G>A,p.Cys634Tyr), consistent with MEN2. Serum calcitonin was 121 pg/ml. She eventually underwent a total thyroidectomy as well.

Genetic testing is beneficial for the diagnosis of symptoms related to genetic pheochromocytoma. It is also beneficial for screening family members and finding tumors at other locations.

KEYWORDS
ectopic ACTH Syndrome, Cushing's syndrome, bilateral pheochromocytoma, medullary thyroid carcinoma, MEN2A