Pheochromocytoma Differs in Patients Under 20

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BETHESDA, MD. — Sustained, severe hypertension was among the clinical features of pheochromocytoma seen more frequently in patients under age 20, compared with adults, Dr. Marta Barontini said at an international symposium on pheochromocytoma sponsored by the National Institutes of Health.

Familial pheochromocytoma—mainly von Hippel-Lindau (VHL) disease—was also more common in the younger patients, "which may account for the noradrenergic profile" of their presenting symptoms, said Dr. Barontini of the center for endocrine research at the R. Gutiérrez Hospital for Children in Buenos Aires.

These findings were based on a review of 58 patients aged 4-20 years—12 boys and 1 girl were younger than 10 years, and the rest were older—who represented 23% of the 255 pheochromocytoma patients studied at the endocrinology research center during a 40-year period. The purpose of the study was to establish the clinical features of pheochromocytoma. Laboratory tests used to make the diagnosis, which was confirmed at the time of surgery, included urinary and plasma catecholamines (epinephrine, norepinephrine, and dopamine), as well as urinary levels of vanillylmandelic acid.

The differences between the clinical signs in the older patients at the center and those in the younger patients were "remarkable," Dr. Barontini said. Sustained hypertension, headaches, and sweating were among the predominant characteristics seen in the younger patients. Fully 93% had severe sustained hypertension, 7% had paroxysmal hypertension, and no patient was normotensive. Of the older patients, 69% had severe sustained hypertension, 26% had paroxysmal hypertension, and 5% were normotensive.

Other clinical signs often found in the younger patients were headaches in 95%, sweating in 90%, blurred vision in 80%, and encephalopathy in 65%. Palpitations, present in 35% of the younger patients, and weight loss, in 15%, were less common than they were in adults, she said.

Among the younger patients, 2% had normal norepinephrine levels, 55% had normal epinephrine levels, and nearly 7% had normal urinary vanillylmandelic acid levels.

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Among patients under age 20, 34% had bilateral adrenal pheochromocytoma and 22% had extra-adrenal pheochromocytoma. Both were higher than they were in older patients.

Among the 58 younger patients, 7 (12%) had a malignant tumor, which was fatal in 4 patients: 1 patient died a few months after surgery and 3 died 8-18 years after surgery. The three patients still alive 5-21 years after surgery include one patient who has hypertension that is treated with four drugs. This patient also has high catecholamine levels and widespread bone metastases, but maintains a good quality of life, Dr. Barontini said.

Familial pheochromocytoma was identified in 36% of those younger than 20 years, compared with 22% of the older patients. Genetic testing, which was performed in familial cases, found some surprising differences, she noted.

VHL disease—an autosomal-dominant neoplasia disorder—had a higher prevalence in the younger population (28%), compared with its incidence in the older patients. Multiple endocrine neoplasia (MEN) type 2a was identified in 2% of the younger patients, MEN type 2b was seen in 2%, neurofibromatosis in 3%, and succinate dehydrogenase subunit B mutations in 2%.

In contrast, the most common mutation identified among the older patients with familial pheochromocytoma was MEN type 2a, in 15%. Dr. Barontini speculated that the higher incidence of VHL in the younger patients may account for the biochemical and clinical features—the noradrenergic profile—seen in this age group. ■

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