PHEOCHROMOCYTOMA

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Annotation: *Pheochromocytomas and paragangliomas are catechol- amine-producing tumors derived rom the sympathetic or parasympathetic nervous system. T ese tumors may arise sporadically or be inherited as eatures o mul- tiple endocrine neoplasia type 2, von Hippel–Lindau disease, or several other pheochromocytoma-associ- ated syndromes. T e diagnosis o pheochromocytomas identi es a potentially correctable cause o hyperten- sion, and their removal can prevent hypertensive crises that can be lethal. T e clinical presentation is variable, ranging rom an adrenal incidentaloma to a hyperten- sive crisis with associated cerebrovascular or cardiac complications.*

EPIDEMIOLOGY

Pheochromocytoma is estimated to occur in 2–8 o 1 million persons per year, and 0.1% o hypertensive patients harbor a pheochromocytoma. T e mean age at diagnosis is 40 years, although the tumors can occur

rom early childhood until late in li e. T e classic “rule o tens” or pheochromocytomas states that 10% are bilateral, 10% are extra-adrenal, and 10% are malignant.

CLINICAL FEATURES

Its clinical presentation is so variable that pheochro- mocytoma has been termed “the great masquerader” (Table 9-1). Among the presenting mani estations, epi- sodes o palpitation, headache, and pro use sweating are typical, and these mani estations constitute a classic triad. T e presence o all three mani estations in asso- ciation with hypertension makes pheochromocytoma a likely diagnosis. However, a pheochromocytoma can be asymptomatic or years, and some tumors grow to a considerable size be ore patients note symptoms.

T e dominant sign is hypertension. Classically, patients have episodic hypertension, but sustained hypertension is also common. Catecholamine crises can lead to heart ailure, pulmonary edema, arrhyth- mias, and intracranial hemorrhage. During episodes o hormone release, which can occur at widely divergent intervals, patients are anxious and pale, and they expe- rience tachycardia and palpitations. T ese paroxysms generally last <1 h and may be precipitated by surgery, positional changes, exercise, pregnancy, urination (par-

ticularly with bladder pheochromocytomas), and vari- ous medications (e.g., tricyclic antidepressants, opiates, metoclopramide).

DIAGNOSIS

T e diagnosis is based on documentation o catechol- amine excess by biochemical testing and localization o the tumor by imaging. T ese two criteria are o equal importance, although measurement o catecholamines

TABLE9-1

PMGrimley:Tumorso theExtra-adrenalParaganglionSystem[Includ- ing Chemoreceptors], Atlas o Tumor Pathology, 2nd Series, Fascicle 9. Washington, DC, AFIP, 1974.) or metanephrines (their methylated metabolites) is tra- ditionally the rst step in diagnosis.

Biochemical testing

Pheochromocytomas and paragangliomas synthe- size and store catecholamines, which include norepi- nephrine (noradrenaline), epinephrine (adrenaline), and dopamine. Elevated plasma and urinary levels o catecholamines and metanephrines orm the corner- stone o diagnosis. T e characteristic uctuations in the hormonal activity o tumors results in consider- able variation in serial catecholamine measurements. However, most tumors continuously leak O-methylated metabolites, which are detected by measurement o metanephrines.

Catecholamines and metanephrines can be measured by di erent methods, including high-per ormance liquid chromatography, enzyme-linked immunosor- bent assay, and liquid chromatography/mass spec- trometry. When pheochromocytoma is suspected on clinical grounds (i.e., when values are three times the upper limit o normal), this diagnosis is highly likely regardless o the assay used. However, as summarized in Table 9-2, the sensitivity and speci city o available biochemical tests vary greatly, and these di erences are important in assessing patients with borderline elevations o di erent compounds. Urinary tests or metanephrines (total or ractionated) and catechol- amines are widely available and are used commonly

CLINICAL FEATURES ASSOCIATED WITH PHEOCHROMOCYTOMA, LISTED BY FREQUENCY OF OCCURRENCE
Headaches
Pro use sweating
Palpitations and tachycardia
Hypertension, sustained or paroxysmal
Anxiety and panic attacks
Pa llo r
Nausea

Abdominalpain Weakness 10. Weight loss 11. Paradoxical response to antihypertensive drugs 12. Polyuria and polydipsia 13. Constipation 14. Orthostatic hypotension 15. Dilated cardiomyopathy 16. Erythrocytosis 17. Elevated blood sugar 18. Hypercalcemia

provocation test, are o relatively low sensitivity and are not recommended.

Diagnostic imaging

A variety o methods have been used to localize pheo- chromocytomas and paragangliomas (able 9-2). C and MRI are similar in sensitivity and should be per ormed with contrast. 2-weighted MRI with gadolinium contrast is optimal or detecting pheo- chromocytomas and is somewhat better than C or imaging extraadrenal pheochromocytomas and para- gangliomas. About 5% o adrenal incidentalomas, which usually are detected by C or MRI, prove to be pheochromocytomas upon endocrinologic evaluation.

umors also can be localized by procedures using

radioactive tracers, including 131I- or 123I-metaiodoben- 111-zylguanidine (MIBG) scintigraphy, In-somatostatin 18 analogue scintigraphy, F-DOPA positron emission tomography (PE), or 18F- uorodeoxyglucose (FDG) PE . Because these agents exhibit selective uptake in paragangliomas, nuclear imaging is particularly use ul in the hereditary syndromes.

DIFFERENTIAL DIAGNOSIS

When the possibility o a pheochromocytoma is being entertained, other disorders to consider include essential hypertension, anxiety attacks, use o cocaine or amphetamines, mastocytosis or carcinoid syndrome (usually without hypertension), intracranial lesions, clonidine withdrawal, autonomic epilepsy, and acti- tious crises (usually rom use o sympathomimetic amines). When an asymptomatic adrenal mass is iden- ti ed, likely diagnoses other than pheochromocytoma include a non unctioning adrenal adenoma, an aldoste- ronoma, and a cortisol-producing adenoma (Cushing’s syndrome).

Conclusion

Thus, the data obtained suggest, as a prevention of DR, the normalization of the following factors: compensation of carbohydrate and lipid metabolism, normalization of blood pressure and, most importantly, the detection of DR at earlier stages for timely and adequate laser coagulation of the retina and surgical treatment.

USED LITERATURES:

1. Dedov I.I., Shestakova M.V. // Soharny diabet - Moscow - 2003 - P. 231-243. 2. Dear T.M. // Diabetic Retinopathy - Moscow – 2000.-P.3.
2. UK Prospektive Diabetes Study (UKPDS) Group: Tight blood pressure control and risk of macrovascular and microvascular complications in type 2 diabetes (UKPDS 38) //
3. BMJ - 1998 - No 317 - P. 703-713.
4. Wilczynski M„ Dziegielewski K. // Klin Oczna - 2005 - No 10-12)-P.672-674.