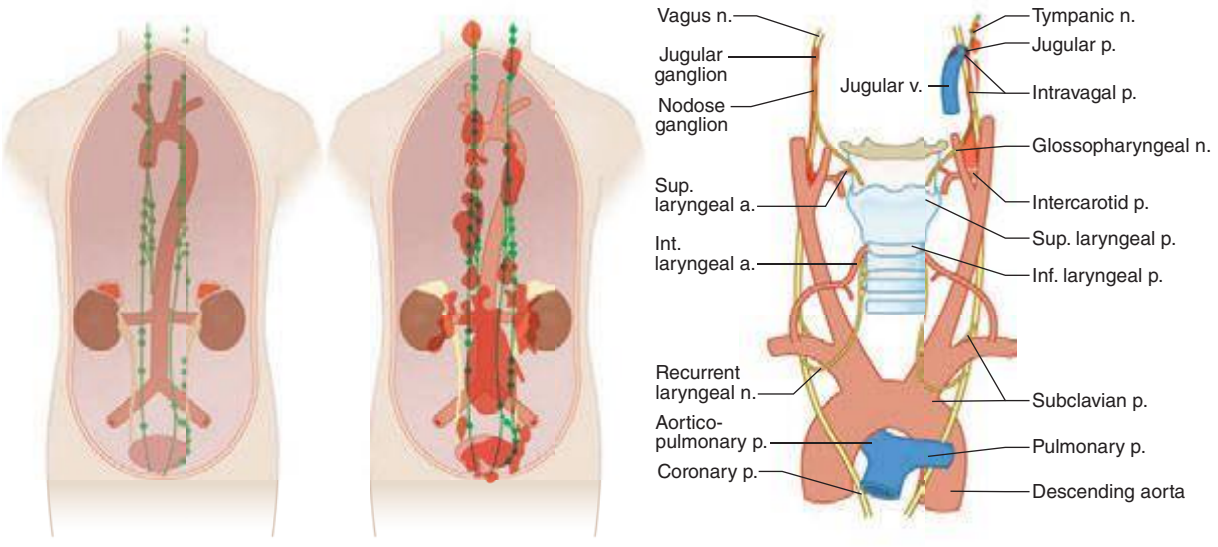

A Adrenal pheochromocytoma

B Extra-adrenal pheochromocytoma

C Head and neck paraganglioma

FIGURE 407-1 The paraganglial system and topographic sites (in red) of pheochromocytomas and paragangliomas. *(Parts A and B from WM Manger, RW Gifford: Clinical and experimental pheochromocytoma. Cambridge, Blackwell Science, 1996; Part C from GG Glenner, PM Grimley: Tumors of the Extra-adrenal Paraganglion System [Including Chemoreceptors], Atlas of Tumor Pathology, 2nd Series, Fascicle 9. Washington, DC, AFIP, 1974.)*

TABLE 407-1 CLINICAL FEATURES ASSOCIATED WITH PHEOCHROMOCYTOMA, LISTED BY FREQUENCY OF OCCURRENCE

1. Headaches
2. Profuse sweating
3. Palpitations and tachycardia
4. Hypertension, sustained or paroxysmal
5. Anxiety and panic attacks
6. Pallor
7. Nausea
8. Abdominal pain
9. 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

TABLE 407-2 BIOCHEMICAL AND IMAGING METHODS USED FOR DIAGNOSIS OF PHEOCHROMOCYTOMA AND PARAGANGLIOMA

Diagnostic Method	Sensitivity	Specificity
24-h urinary tests		
Catecholamines	+++	+++
Fractionated metanephrines	++++	++
Total metanephrines	+++	++++
Plasma tests		
Catecholamines	+++	++
Free metanephrines	++++	+++
Imaging		
CT	++++	+++
MRI	++++	+++
MIBG scintigraphy	+++	++++
Somatostatin receptor scintigraphy[a]	++	++
^{18}Fluoro-DOPA PET/CT	+++	++++

[a]Values are particularly high in head and neck paragangliomas.

Abbreviations: MIBG, metaiodobenzylguanidine; PET/CT, positron emission tomography plus CT. For the biochemical tests, the ratings correspond globally to sensitivity and specificity rates as follows: ++, <85%; +++, 85–95%; and ++++, >95%.

this circumstance, it is important to exclude dietary or drug-related factors (withdrawal of levodopa or use of sympathomimetics, diuretics, tricyclic antidepressants, alpha and beta blockers) that might cause false-positive results and then to repeat testing or perform a clonidine suppression test (i.e., the measurement of plasma normetanephrine 3 h after oral administration of 300 μg of clonidine). Other pharmacologic tests, such as the phentolamine test and the glucagon provocation test, are of relatively low sensitivity and are not recommended.

Diagnostic Imaging A variety of methods have been used to localize pheochromocytomas and paragangliomas (Table 407-2). CT and MRI are similar in sensitivity and should be performed with contrast. T2-weighted MRI with gadolinium contrast is optimal for detecting pheochromocytomas and is somewhat better than CT for imaging extraadrenal pheochromocytomas and paragangliomas. About 5% of adrenal incidentalomas, which usually are detected by CT or MRI, prove to be pheochromocytomas upon endocrinologic evaluation.

Tumors also can be localized by procedures using radioactive tracers, including ^{131}I- or ^{123}I-metaiodobenzylguanidine (MIBG) scintigraphy, ^{111}In-somatostatin analogue scintigraphy, ^{18}F-DOPA positron emission tomography (PET), or ^{18}F-fluorodeoxyglucose (FDG) PET. Because these agents exhibit selective uptake in paragangliomas, nuclear imaging is particularly useful in the hereditary syndromes.

Differential Diagnosis When the possibility of a pheochromocytoma is being entertained, other disorders to consider include essential hypertension, anxiety attacks, use of cocaine or amphetamines, mastocytosis or carcinoid syndrome (usually without hypertension), intracranial lesions, clonidine withdrawal, autonomic epilepsy, and factitious crises (usually from use of sympathomimetic amines). When an asymptomatic adrenal mass is identified, likely diagnoses other than pheochromocytoma include a nonfunctioning adrenal adenoma, an aldosteronoma, and a cortisol-producing adenoma (Cushing's syndrome).

TREATMENT PHEOCHROMOCYTOMA

Complete tumor removal, the ultimate therapeutic goal, can be achieved by partial or total adrenalectomy. It is important to preserve the normal adrenal cortex, particularly in hereditary disorders in which bilateral pheochromocytomas are most likely. Preoperative preparation of the patient is important. Before surgery, blood pressure should be consistently below 160/90 mmHg. Classically, blood pressure has been controlled by α-adrenergic blockers (oral