Answer

Paraganglioma

The metaiodobenzylguanidine scan (Figure 1) shows increased uptake in a small region adjacent to the junction of the abdominal aorta and the inferior mesenteric artery; computed tomographic scan (Figure 2) reveals a 2.5-cm mass in the same region. The preoperative diagnosis of paraganglioma was made and the patient opted for surgical resection of the mass. The patient underwent preoperative preparation by fluid volume expansion and α-blockade with phenoxybenzamine hydrochloride. An exploratory laparotomy was performed and the mass was dissected off the inferior mesenteric artery and aorta. A complete oncological resection was obtained. Pathology revealed evidence of positive staining of neuron-specific enolase, synaptophysin, chromogranin, and S100. His blood pressure normalized after the operation.

Paragangliomas, also known as extra-adrenal pheochromocytomas, are rare catecholamine-releasing neuroendocrine tumors. As with intra-adrenal pheochromocytomas, patients often present with nonspecific symptoms related to catecholamine excess, including episodic headache (80%), palpitations (64%), and diaphoresis (57%). This triad in a patient with hypertension has 90.9% sensitivity and 93.8% specificity for pheochromocytoma.[1] In a prior study, 56% also experienced nausea and vomiting, 35% exhibited abdominal pain, and 8% noted constipation.[2] Tumor size is positively correlated with secretory activity, with larger tumors usually secreting high levels of catecholamines.[3]

The vast majority of pheochromocytomas are located within the adrenal gland, but approximately 10% to 15% are extra-adrenal paragangliomas. Paragangliomas almost exclusively occur in the abdomen (98%), with the organ of Zuckerkandl being the predominant location.[4] Functional secretory tumors make up 77% of paragangliomas of the organ of Zuckerkandl. Paragangliomas are of particular importance because of their rates of malignancy and recurrence. While approximately 10% of intra-adrenal pheochromocytomas are malignant, paragangliomas have a rate of malignancy approaching 50%.[5,6]

The preferred biochemical test for paragangliomas is the measurement of free plasma metanephrine levels (sensitivity 99%).[7-9] Biopsy is contraindicated for suspected pheochromocytomas and paragangliomas because of the high rate of complications.[10] Intra-adrenal pheochromocytomas generally show elevated plasma epinephrine levels while extra-adrenal paragangliomas generally produce elevated norepinephrine levels, although 10% of paragangliomas of the organ of Zuckerkandl produce epinephrine.[6] By itself, metaiodobenzylguanidine scintigraphy is 80% to 90% sensitive in detecting paragangliomas,[11] while computed tomography and magnetic resonance imaging are 95% and nearly 100% sensitive,[6] respectively. Thus, if a pheochromocytoma is biochemically confirmed, the ideal algorithm is to conduct a metaiodobenzylguanidine scan to determine the location of the tumor, followed by high-resolution computed tomography or magnetic resonance imaging to determine proximity to and invasion of neighboring structures.

Surgical resection is the treatment of choice. Although laparoscopic adrenalectomy is the standard for intra-adrenal pheochromocytomas, paragangliomas often require open procedures because of the difficulty of the dissection and invasion into vital neighboring structures. Preoperative preparation via α-blockade with phenoxybenzamine is advised to prevent hypertensive crisis. Volume expansion via intravenous fluids is recommended to reduce the likelihood of cardiovascular collapse on removal of the tumor.

Lifelong follow-up of patients with paraganglioma is advised because recurrence of disease can occur decades after successful surgical resection.[7] The 5-year survival of patients with paraganglioma after successful surgical resection is 75%. However, the 10-year disease-free survival is only 45%, suggesting that there is a strong likelihood for recurrence in more than half of patients successfully treated surgically.[12]

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