Transformation of a Nonfunctional Paraganglioma With I-123 MIBG Scintigraphy Correlation

A Case Report

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Abstract: A 35-year-old woman presenting with abdominal pain was found to have mildly elevated catecholamine levels and a retroperitoneal mass. The patient underwent a negative I-123 MIBG scintigraphy scan and a nondiagnostic fine needle aspiration. Eleven years later the patient presented with a hypertensive emergency and markedly elevated catecholamine levels. A subsequent I-123 MIBG scintigraphy scan showed intense uptake corresponding to the previously seen retroperitoneal mass. The patient underwent surgical resection and pathology confirmed the presence of a paraganglioma.

A paraganglioma is an extra-adrenal pheochromocytoma that contains chromaffin cells and is thus capable of producing catecholamines. I-123 metaiodobenzylguanidine (MIBG) scintigraphy has become the imaging study of choice for paragangliomas and has a sensitivity of ~77% to 100% in detecting functional paragangliomas.1 This case demonstrates scintigraphic correlation of the functional transformation of a nonfunctional paraganglioma in a time span of ~10 years. Although there are previously published case reports of scintigraphic positive, nonfunctional paragangliomas2,3 and scintigraphic negative chromaffin cell tumors,4,5 there has been no prior documented case of scintigraphic transformation on MIBG.

Abbreviations: CT = computed tomography, DOPA = dopamine, DTPA = diethylenetriamine pentaacetate, FDG = fluorodeoxyglucose, MIBG = metaiodobenzylguanidine.

CASE PRESENTATION

A 35-year-old woman presented to an emergency department with abdominal pain, nausea, and vomiting. A computed tomography (CT) scan was performed (unavailable) and she was diagnosed with gallstones. Her CT scan also revealed a 4 × 3 cm heterogeneous mass within the right upper quadrant, below the third portion of the duodenum. The origin of the mass was unclear and tumors of the small bowel, pancreas, and adrenal gland were all considerations at this time. The patient was admitted for pain control and additional work-up of the right upper quadrant mass. Her work-up included a fine needle aspiration of the mass, but this was unfortunately nondiagnostic. The patient’s pain subsided after a few days and she was discharged due to a desire to be home for the Thanksgiving holiday. The patient presented to our nuclear medicine department after the holiday to undergo an MIBG study (Figure 1). The patient never received any treatment for the mass and was subsequently lost to follow-up.

Eleven years after the initial work-up, the patient returned to our institution after suffering an episode of severe epigastric pain, headache, and acute loss of consciousness. Her urine catecholamines (norepinephrine and vanillylmandelic acid) were found to be markedly elevated: epinephrine–7 (0–15 mcg/24 h), norepinephrine–1289 (11–86 mcg/24 h), and vanillylmandelic acid–29.5 5 (2–10 mg/24 h). Of note, her urine catecholamines 11 years earlier were basically within normal limits. The patient underwent surgical resection and pathology confirmed the presence of a paraganglioma.
FIGURE 2. Axial (A) and coronal (B), contrast-enhanced CT of the abdomen/pelvis demonstrating a large, heterogeneous mass containing solid and cystic components within the right retroperitoneum (white arrow). Incidentally noted large, fibroid uterus (black arrow). Axial, T2-weighted fat-saturation magnetic resonance imaging (MRI) (C) demonstrating a complex mass within the right retroperitoneum (white arrow). Note the relative hyperintensity of the cystic components within the mass. Axial, T1-weighted MRI (D) demonstrates a hypointense mass within the right retroperitoneum. CT = computed tomography.

FIGURE 3. I-123 MIBG scintigraphy showing intense uptake corresponding to the right retroperitoneal mass at 6 h (white arrows) and 24 h (black arrows) after infusion. MIBG = metaiodobenzylguanidine.
normal limits: epinephrine–18, norepinephrine–91, and vanillylmandelic acid–4. Another CT scan was performed and this time revealed a 7 × 5 cm mass in the right upper quadrant, below the third portion of the duodenum (Figure 2). Subsequent evaluation with I-123 MIBG scintigraphy revealed markedly increased uptake within the right upper quadrant mass (Figure 3). The patient underwent surgical resection and pathology confirmed the presence of a paraganglioma (Figure 4).

The patient was unreachable to discuss consent for publication.

DISCUSSION

I-123 MIBG will concentrate by active transport in catecholamine-producing paragangliomas. At the time of the patient’s initial presentation, we believe the I-123 MIBG scan was negative due to a nonfunctioning paraganglioma that eventually transformed to a functioning paraganglioma. This scenario is also corroborated by the marked elevation in catecholamine levels that occurred between the patient’s initial and secondary presentations. Another possibility that could have explained the initially negative MIBG scan would be poor patient preparation; MIBG uptake can be inhibited by any drug that compromises transport into adrenergic tissue, such as antihypertensives, antidepressants, or antipsychotics. However, a review of the patient’s chart and medications revealed no history of such medications.

The overall sensitivity and specificity of I-123 MIBG for the detection of paragangliomas is 56% to 76% and 84% to 100%, respectively. However, it has been reported that the sensitivity of I-123 MIBG decreases when evaluating for metastatic paragangliomas, it is conceivable that alterations in gene expression over time could also be responsible for the development of a functional paraganglioma in our case. To our knowledge, this is the first reported case of the transformation of nonfunctioning paraganglioma to a functioning paraganglioma.

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