

Dopamine-Producing Paraganglioma Not Visualized by Iodine-123-MIBG Scintigraphy

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A patient with an aorticopulmonary paraganglioma was found to have normal plasma norepinephrine and epinephrine levels and elevated dopamine levels. Iodine-123-MIBG scintigraphy did not visualize this tumor. The same finding on a negative MIBG scan in two patients with exclusively dopamine-secreting chemodectomas has been previously reported. In our patient, [^{111}In -DTPA-D-Phe1]-octreotide scintigraphy proved to be an effective imaging technique.

Key Words: dopamine; iodine-123-MIBG scintigraphy; paraganglioma; chemodectoma

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Extra-adrenal paragangliomas are tumors with catecholamine-producing potential. Intrathoracic paragangliomas most frequently arise in the paraganglia anatomically related to the pulmonary artery and aortic arch. Most functionally active paragangliomas secrete norepinephrine or norepinephrine with only small amounts of epinephrine (1). Exclusive or predominant dopamine secretion by paragangliomas has only occasionally been reported (2,3).

CASE REPORT

In March 1992, a 54-yr-old female was referred to a pulmonologist because of dyspnea and constant retrosternal pain. Except for obesity (height 155 cm and weight 79 kg), her physical examination was unremarkable. Her blood pressure was 160/95 mmHg and her heart rate was 68 bpm. A chest x-ray showed an elevation of the right hemidiaphragm and a mass in the left hilum in the aortic window. During a sniff test, the right hemidiaphragm seemed to move paradoxically. Roentgenologic examination of the cervical spine revealed no abnormalities. Subsequent CT of the thorax showed a mass (diameter 3 cm) next to the aorta and superior to the left pulmonary artery (Fig. 1). Plasma catecholamine measurements were repeatedly performed and showed normal epinephrine and norepinephrine but elevated dopamine levels (Table 1). Iodine-123-MIBG scintigraphy was negative, whereas [^{111}In -DTPA-D-Phe1]-octreotide (pentetreotide) scintigraphy

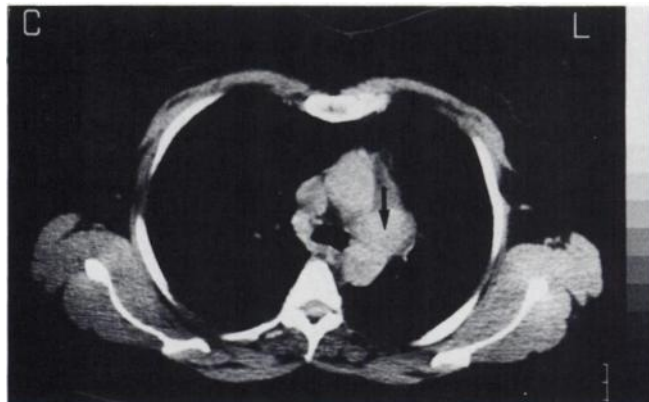


FIGURE 1. CT scan of the thorax shows a mass adjacent to the inside bend of the aortic arch.

clearly showed a hot spot in the left hilar region (Fig. 2). In view of the fact that surgical removal of the tumor in this obese patient, with a paralyzed hemidiaphragm, could potentially damage the left phrenic nerve and subsequently lead to respiratory insufficiency, surgical treatment was postponed until considerable weight loss had been achieved. Thoracotomy was performed in December 1992. A tumor was found adjacent to the inside bend of the aortic arch near ligamentum arteriosum. The phrenic and recurrent nerves were identified and spared and the tumor was resected in toto. Regional lymph nodes were free of tumor. Recovery was prompt and the patient left the hospital 9 days after the operation. In May 1993, plasma catecholamine measurements were obtained and dopamine levels appeared to be normalized (Table 1).

DISCUSSION

The paraganglioma in this patient originated from the paraganglionic cells comprising aortic body chemoreceptors. These aorticopulmonary paragangliomas usually are asymptomatic at presentation and are often discovered incidentally on a chest roentgenogram obtained for other reasons (4). Computed tomography (CT) is the most commonly used imaging technique (5), whereas ^{123}I -MIBG scintigraphy is used to assess the presence of extra-adrenal or metastatic localizations (6). Pentetreotide scintigraphy, on the other hand, is an excellent alternative imaging technique for the localization of paragangliomas (7-9).

Plasma catecholamine measurements in our patient

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TABLE 1
Plasma Catecholamine Measurements

	Preoperative		Postoperative
	March 3, 1992	April 23, 1992	May 27, 1993
Norepinephrine (pg/ml)	280	172	337
Epinephrine (pg/ml)	50	42	45
Dopamine (pg/ml)	303	431	28

The range of normal values of plasma norepinephrine, epinephrine and dopamine in our laboratory is, respectively, 100–500 pg/ml, 10–100 pg/ml and 5–120 pg/ml.

showed an isolated elevation of dopamine. The exclusive secretion of dopamine by paragangliomas is unusual (1,10). In a series of 50 patients with paraganglioma, Proye et al. (11) found only two tumors that exclusively produced dopamine. They concluded that these two tumors, as did the tumor in our patient, did not take up ¹³¹I-MIBG, whereas the other tumors did. Cellular uptake of ¹²³I-MIBG occurs by a specific (uptake-one) and a nonspecific uptake system (12). After uptake into the cell, ¹²³I-MIBG is stored within neurosecretory granules (13). Usually these granules are abundantly present within the cytoplasm of paraganglioma cells. A positive association between the semiquantitatively determined amount of neurosecretory granules and the percentage uptake of radioactivity per gram of tumor tissue has been demonstrated for paragangliomas (14). Electron microscopy studies performed on the tumor of our patient showed a total absence of cytoplasmic neurosecretory granules (Fig. 3). This may explain why the ¹²³I-MIBG scan was negative. The positive experience with pentetretotide scintigraphy as opposed to MIBG scintigraphy suggests that this technique may be the diagnostic procedure of first choice to detect paragangliomas.

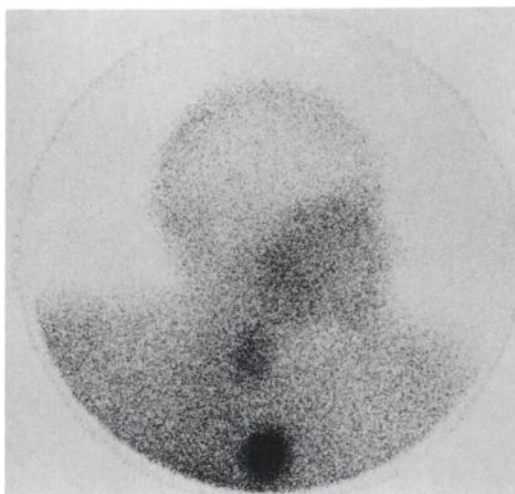


FIGURE 2. Pentetretotide scintigraph of the right lateral part of head and upper part of the chest (anterior view) shows an intrathoracic hot spot under the usual thyroid gland activity.

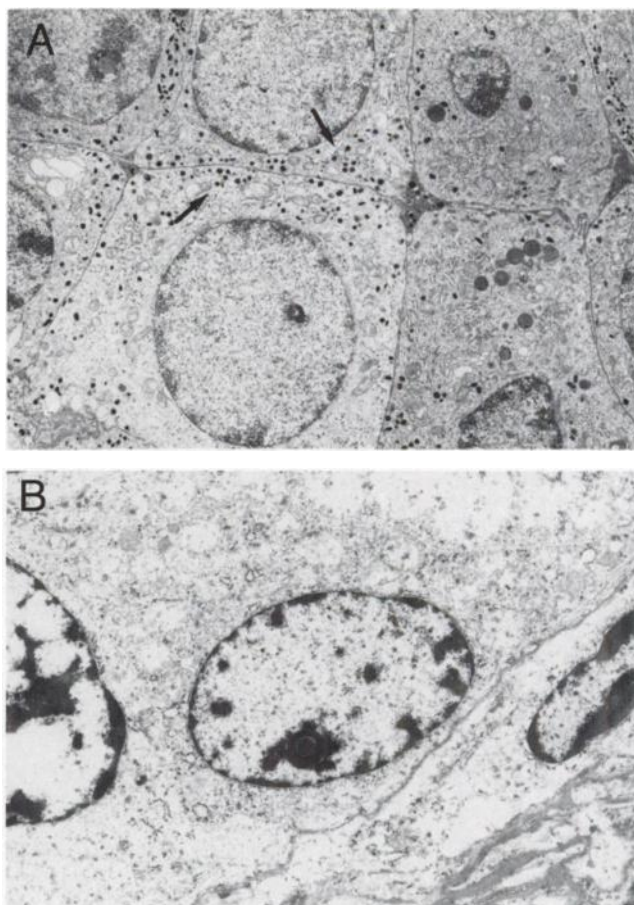


FIGURE 3. (A) EM photograph of a paraganglioma shows neurosecretory granules within the cytoplasm and (B) EM photograph of the tumor in our patient. Notice the absence of neurosecretory granules.

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