Localization of Mediastinal Paragangliomas (Pheochromocytoma)*

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While the incidence of catecholamine-producing tumor is only 1 to 2 per 100,000 adults per year, the clinical presentation is often an exciting one and surgery offers the only cure. The evolution of more specific catecholamine measurements has made the diagnosis of a catecholamine-producing tumor much easier and CT scanning can identify those tumors located in the adrenal medulla (pheochromocytoma) in over 90 percent of cases.* However, extraadrenal primary catecholamine-producing tumors (paragangliomas) and metastatic pheochromocytomas are often undetected by CT scanning until they are relatively large compared to those within the adrenal gland, and their localization remains a challenge. These extraadrenal tumors may account for up to 19 percent of catecholamine-producing tumors; most are in the abdomen, but about 2 percent are within the chest.

Within the thorax, primary paragangliomas can arise along the sympathetic ganglia and at the site of the aortic and pulmonary bodies. The tumors are usually mediastinal and paravertebral in location, are more common in males, (2:1) and one third occur in children. Eight percent are malignant and can be locally invasive. These tumors can be multiple and at the site of paraganglion tissue, as exemplified by a case with over 21 primary tumors in the neck, chest, and abdomen. More recently, primary cardiac paragangliomas arising from the atrial wall and the atrial septum have been described.8,9

The localization of these mediastinal and cardiac paragangliomas has been greatly aided by two recently developed techniques: ¹¹I-MIBG scintigraphy and two-dimensional echocardiography. These techniques have permitted more direct CT scanning, including reconstructions to more precisely define the anatomy of the tumor. ¹¹I-metaiodobenzylguanidine (¹¹I-MIBG) is a new radiopharmaceutical developed at the University of Michigan that has proven to be very useful in the localization of catecholamine-producing tumors, neoroblastomas and carcinoid tumors.¹⁰ MIBG is structurally similar to norepinephrine, guanethidine and guanadrel and is taken up and concentrated in adrenergic vesicles. Drugs known to interfere with norepinephrine uptake such as reserpine, some antidepressants, sympathomimetic drugs and antihistamines will block MIBG uptake as well. Catecholamine-producing tumors show up as areas of intense uptake (Fig IA-D). In our evaluation of 48 patients, there were 20 true positives (six pheochromocytomas, four paragangliomas, ten metastatic or recurrent pheochromocytomas) and 24 true negative studies. There was one false positive study near the bladder (CT scan also falsely positive) and six false negative studies (three pheochromocytomas, one paraganglioma, two metastatic disease). The overall sensitivity was 77 percent, specificity 96 percent, and accuracy 86 percent.¹¹ While the advantage of this technique is the ease of scanning the entire body, there are more false negatives than with CT. Although the exact anatomic location cannot be ascertained from scintigraphic studies, such studies can direct subsequent CT scanning to the area of interest.¹² ¹¹I-MIBG is of particular value in assessment of extra adrenal paraganglioma, familial pheochromocytoma, recurrence or malignant pheochromocytoma. ¹¹I-MIBG is also being studied in the treatment of inoperable pheochromocytoma. Some early results provide guarded optimism as to its final place in the treatment of this tumor which typically is resistant to roentgen therapy and chemotherapy.¹³

Cueto-Garcia et al have recently reported the use of two-dimensional echocardiography utilizing the suprasternal approach to detect a paraganglioma in the aortico-pulmonary region of the middle mediastinum (see page 834). The anatomic relationships to other vascular structures in the mediastinum were delineated as well. The location and nature of the tumor were further defined by ¹¹I-MIBG scintigraphy and com-
These three imaging techniques ($^{123}$I-MIBG, 2-D echocardiography, CT scan) have greatly increased the ability to localize intrathoracic catecholamine-producing tumors. None is 100% specific, accurate and sensitive, but they complement each other in the search for these tumors. Their use is suggested when the biochemical diagnosis of a catecholamine-producing tumor is secure and CT imaging within the abdomen is negative. These tumors now should be localized with greater ease and permit early operation so that the cure rate for extraadrenal tumors can be expected to improve.

REFERENCES

Localization of Mediastinal Paragangliomas (Sheps, Brown)
33rd Annual James J. Waring Chest Conference

The American Lung Association of Colorado, North Central Region, will present this course August 29-31 in Estes Park, Colorado. For information, contact Ms. Shirley Lindquist, Regional Director, Colorado Trudeau Society, PO Box 921, Loveland 80539 (303) 667-5198.

Allergy and Clinical Immunology

The Association for the Care of Asthma, and Children's Hospital National Medical Center, will present this course on Mackinac Island, Michigan, July 16-18. For information, contact Mr. Harold A. Ifft, Executive Director, Association for the Care of Asthma, Box 565, Spring Valley Road, Ossining, New York 10562 (914) 762-1941.