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# CARNEY'S TRIAD PARAGANGLIOMAS

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Carney's triad is the association of gastric stromal sarcoma, pulmonary chondroma, and extra-adrenal paraganglioma.[1] Paragangliomas in Carney's triad are frequently multiple and arise from the mediastinum, aortopulmonary body, or thoracic sympathetic chain.[2] Key surgical features of all paragangliomas are their intense vascularity, ability to produce catecholamines, and potential growth into vital structures. We describe the technically challenging resection of a vascular mediastinal paraganglioma in a patient with Carney's triad.

**Clinical summary.** A 59-year-old man was seen by us in February 1999 with tremors, intermittent flushing, and anxiety. He and his 2 brothers had undergone resection of bilateral carotid paragangliomas in 1989. Now, 10 years later, a diagnostic workup revealed elevated urine vanillymandelic acid. Computed tomography (CT) of the abdomen to rule out adrenal disease was remarkable for a submucosal exophytic mass (5.4 × 7.1 cm) in the stomach. The patient underwent subtotal distal gastrectomy for what was revealed to be a gastrointestinal stromal tumor. On follow-up, chest CT revealed a mediastinal tumor (7.5 × 4.5 cm) contiguous with the aorta and pulmonary artery (Fig 1). Tests for serum and urinary catecholamines were negative. Nuclear medicine 111-OctreoScan (Mallinckrodt, St Louis, Mo) showed focally increased uptake in the midportion of the middle mediastinum (Fig 2). There was no uptake in the neck or adrenals.

The patient underwent surgery once again. A median sternotomy revealed tumor completely filling the pretracheal compartment and subaortic space with projection into the aortopulmonary window. The tumor was extremely vascular; an estimated 3.7 L of blood was lost in the course of mobilization. Despite this, complete resection was accomplished.

**Discussion.** Carney's triad consists of 3 unusual tumors: gastric stromal sarcoma, pulmonary chondroma, and extra-adrenal paraganglioma (chemodectoma, carotid body tumor).[1] A total of 79 cases of this triad have been reported in the literature, the vast majority with 2 or 3 tumors. Despite the malignant nature of these tumors, most are indolent.[1]

Paragangliomas in Carney's triad arise from paraganglia tissue dispersed in the chest, abdomen, head, and neck. Histologically, cells are arranged in a trabeculoalveolar pattern with highly vascular fibrous septa. Our patient had bilateral carotid body tumors and a third paraganglioma in the mediastinum. Except in patients with Carney's triad, the mediastinum is a very unusual site for paragangliomas.[2] A high incidence of aggressive growth has been reported in mediastinal paragangliomas with invasion of the great vessels.[1] Therefore, early diagnosis is important.

Patients with paragangliomas often have a mass or signs of catecholamine excess. Mediastinal paragangliomas may also cause symptomatic compression of the great vessels or of the recurrent laryngeal nerve. Diagnostic workup should include evaluation of urinary catecholamine metabolites. Localization is through chest radiograms, CT, and metaiodobenzylguanidine (MIBG) scintigraphy. However, for detecting paragangliomas, In 111 octreotide acetate scintigraphy (Sandostatin; Sandoz Pharmaceuticals, East Hanover, NJ) has been advocated as superior to MIBG scintigraphy.[3,4] In a study directly comparing the 2 methods in 8 patients with histologically confirmed paragangliomas, the authors reported a sensitivity of 50% for MIBG and 100% for In 111 octreotide scintigraphy using In 111 chloride–labeled octreotide.[3] In another study, Kwekkeboom and associates[4] used In 111 octreotide scintigraphy to identify 50 of 53 known paragangliomas (sensitivity 94%); 9 additional paragangliomas, not detected by routine methods, were identified by means of the whole-body

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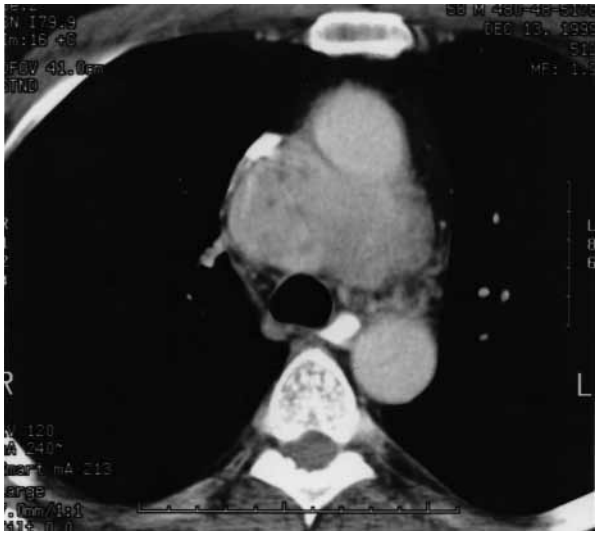

**Fig 1.** Chest CT scan showing a large mediastinal tumor contiguous with the aorta and pulmonary artery.

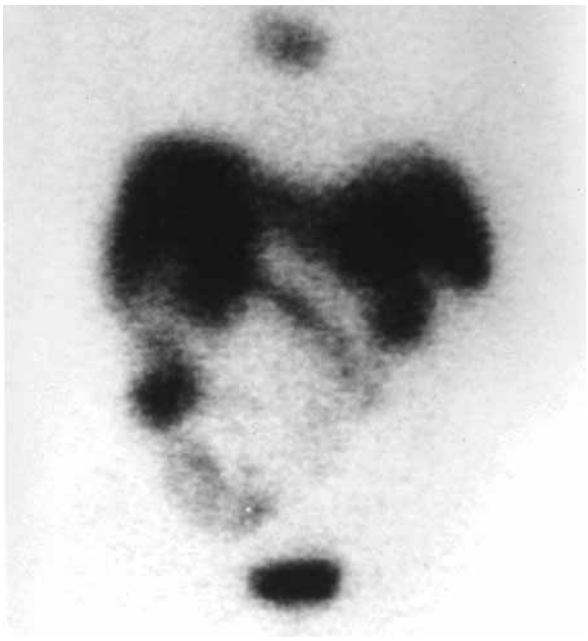

**Fig 2.** OctreoScan shows abnormally increased uptake in the mediastinum. The liver, spleen, and kidneys normally accumulate radioactivity.

octreotide scan.[4] For this reason, In 111 octreotide scintigraphy may be particularly useful in patients with Carney's triad given the multiplicity of the paragangliomas. We, therefore, used octreotide scintigraphy to confirm our diagnosis of mediastinal paraganglioma and to rule out concomitant neck and abdominal foci of disease.

Because of the radioresistance of paragangliomas, surgical resection is the treatment of choice.[2] These tumors are highly vascular and often adhere to vital structures. Therefore, preoperative embolization of tumors has been advocated to decrease technical difficulty, blood loss, and operative time.[5] The mediastinal tumor resected in our patient was highly vascular and friable, shedding blood with minimal perturbation. Almost 4 L of blood was lost. This high blood loss heightened the difficulty of an already treacherous dissection to separate the tumor from the aorta and pulmonary artery. Preoperative embolization would have been very useful to decrease the size and vascularity of the tumor.

Finally, Carney's triad and familial paraganglioma may be genetically linked. Three patients in the literature with this triad have siblings with bilateral paragangliomas, in accordance with the Mendelian inheritance pattern observed in familial paragangliomas.[1] This unusual occurrence is not likely due to chance.

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