A 50-year-old woman was admitted with angina. Her past medical history included poorly controlled hypertension and mild Parkinson’s disease. A systemic examination was unremarkable apart from a slight resting tremor. Serial ECGs and 12-hour postadmission troponin levels were normal. Echocardiography confirmed normal biventricular function and no left ventricular hypertrophy. A 12-lead exercise stress ECG was stopped at stage 2 of the Bruce protocol because of shortness of breath. The patient had achieved 82% of her target heart rate with no ischemic ECG changes.

Selective coronary angiography was performed and showed normal left anterior descending and circumflex arteries. However, the right ventricular branch of the right coronary artery appeared to supply a vascular mass (Figure 1 and Data Supplement Movie I). Subsequent computed tomography and magnetic resonance imaging demonstrated an intrapericardial mass with compression of the right pulmonary vein and the posterior wall of the superior vena cava (Figures 2 and 3). There was no evidence of metastases. Her 24-hour urine norepinephrine level was elevated to 344.6 μg/d (normal, 10 to 150 μg/d), with normal epinephrine and dopamine levels measured at 6 μg/d and 201 μg/d (normal values <12 μg/d and 130 to 1200 μg/d), respectively.

Surgical intervention was planned, and a multinodular encapsulated intrapericardial tumor with a gray-tan appearance measuring 2.5×3.0×4.0 cm was resected. There was no evidence of any intracardiac extension. Histological analysis of the tumor confirmed a compact nested appearance (the German expression is *zellballen*, meaning balls of cells), giving rise to a pseudoglandular pattern (Figure 4). The cells displayed a granular eosinophilic cytoplasm, central nuclei, and prominent nucleoli with no mitotic features. Immunohistochemistry was strongly positive for chromogranin A (Figure 5) and synaptophysin in the tumor cells. S-100 staining was highly positive in the surrounding sustentacular cells (Figure 6) and staining for cytokeratin was negative, thereby...
excluding a carcinoid tumor. The histological and clinical appearances were those of an intrapericardial paraganglioma.

The patient recovered uneventfully from surgery and maintains excellent blood pressure control with bendrofluazide. Subsequent 24-hour urine collection 22 months after surgery demonstrated normal norepinephrine, epinephrine, and dopamine excretion.

Catecholamine-secreting tumors arise from the neural-crest cells and have an annual incidence of 1 to 2 per 100 000. Eighty percent arise within the adrenal glands (pheochromocytomas), and the remaining 20% are extra-adrenal (paragangliomas).1 Two percent of paragangliomas are within the chest. These may originate within the mediastinum, lungs, heart, or esophagus. The majority of the mediastinal paragangliomas are found in the anterior or posterior aortopulmonary groove and are largely nonfunctioning.2 Functioning intrapericardial paragangliomas are exceedingly rare. More than 90% of paragangliomas are benign and compress adjacent structures as they enlarge, resulting in chest pain, dyspnea, cough, dysphagia, or hoarseness. However, a minority are locally invasive and exhibit distal metastases.3 Echocardiography, computed tomography, and magnetic resonance imaging help localize a paraganglioma.131I-metaiodobenzylguanidine scintigraphy shows paragangliomas as areas of intense uptake. Plasma and urinary catecholamine levels may be raised if a tumor is functioning. Complete resection of the tumor is curative, although ligation of the feeding vessels leading to infarction in a surgically unresectable tumor has been described in the literature.4 Histological and immunohistochemical analyses are invaluable diagnostic adjuncts in establishing the diagnosis.

Disclosures

None.

References

Figure 5. Immunohistochemical staining with chromogranin A is strongly positive in the tumor cells. Magnification ×200.

Figure 6. Immunohistochemical staining with S-100 is intensively positive in the sustentacular cells (arrows). Magnification ×200.
Intrapericardial Paraganglioma Presenting as Chest Pain
Omar Rana, Peter Gonda, Bruce Addis and Kim Greaves

Circulation. 2009;119:e373-e375
doi: 10.1161/CIRCULATIONAHA.108.822197
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2009 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/119/12/e373

Data Supplement (unedited) at:
http://circ.ahajournals.org/content/suppl/2009/04/01/119.12.e373.DC1

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/