is an important determinant of arterial elasticity and it is generally accepted that arterial elasticity tends to decrease with age.14–17

In conclusion, MRI enables the assessment of aortic elasticity in patients with Marfan’s syndrome with elective ARR. Aortic elasticity between Marfan patients with and without aortic root replacement showed no differences.


Echocardiographic Features of Pheochromocytoma of the Heart

Martin Osranek, MD, Francesca Bursi, MD, George M. Gura, MD, William F. Young, Jr., MD, and James B. Seward, MD

Primary cardiac pheochromocytomas are extremely rare.1 The first case was reported in 1974 by Besterman et al;2 a total of 30 cases were reported before 1992,3 and another 20 were reported in the subsequent 10 years.3–6 The 2-dimensional echocardiograms of Mayo Clinic patients with primary cardiac pheochromocytoma were reviewed. We noted a characteristic appearance of this rare tumor that has been unappreciated.

A complete search of the Mayo Clinic echocardiographic and clinical database identified 8 patients with primary pheochromocytoma of the heart. Echocardiography consistently identified characteristic tumor locations, size, shape, and consistency. Transesophageal echocardiography (TEE) uniformly gave better delineation of the characteristic features. All tumors were located along the atrioventricular groove, in the distribution of the epicardial coronary arteries (Figures 1 to 3). All tumors were well circumscribed and ovoid, ranging from 1.5 to 5.1 cm in diameter, with a fine granular appearance. The primary epicardial coronary artery was immediately adjacent to the tumor; however, the coronary artery was typically not enveloped by the tumor. A distinct tumor capsule was not apparent.

The average age at diagnosis was 38 years (range 25 to 57), and most of the patients were women (75%). TEE most consistently showed characteristic tumor appearance. Confirmatory imaging and diagnosis included computed tomography, magnetic resonance imaging, octreotide scintigraphy, and total body...
iodine I-131 metaiodobenzylguanidine (MIBG) scintigraphy. However, transthoracic echocardiography, computed tomography, and MIBG scintigraphy showed isolated false-negative results (Table 1). When coronary angiography was performed, it uniformly showed a coronary neovascular blood supply. All patients had increased blood or urine levels of catecholamines or their metabolites.

In this series of patients with primary pheochromocytoma of the heart, all tumors shared a characteristic echocardiographic appearance that, to date, has not been seen with any other tumor type. TEE consistently delineated these features in all of our patients. Recognition of these features proved beneficial, heightening awareness and prompting further confirmation. Angiographic media can precipitate a hypertensive crisis; therefore, preprocedural diagnosis can avoid potential risk to the patient.

Cardiac pheochromocytomas typically occur in young patients and are occasionally familial or multiple as part of multiple paraganglioma syndrome. Cardiac pheochromocytomas are usually benign. A syndrome (the Carney triad) comprising functional extra-adrenal catecholamine-producing tumor, gastric epithelioid leiomyosarcoma, and pulmonary chondroma was present in 1 of our patients (case 7).

Pheochromocytomas are homogeneous, well demarcated but not encapsulated, and highly vascularized tumors consisting of paraganglionic tissue. The heart shows the typical Zellballen growth pattern and positive staining for chromagranin and neuron-specific enolase. Pathologic descriptions of primary cardiac pheochromocytomas confirm their location on the surface of the heart adjacent to the coronary arteries, typically near the origin of the great arteries, in the atrioventricular grooves, in the interatrial septum, and protruding into the atria. The consistency of the locations can be partly explained by the normal occurrence of microparaganglia in the atrial and aortocoronary regions. Tumors receiving coronary blood supply show a constant microanatomic feature that has the specific morphologic characteristics of pressor-sensing vessels. Glenner and Grimley proposed a classification according to which coronary and aortocopulmonary paragangliomas arise from branchial arch-derived structures.

Pheochromocytomas mainly come to clinical attention because of symptoms related to their catecholamine secretion. There are case reports of patients with ischemic heart disease caused by compression of the left main coronary artery and compressive dysphagia. Compared with adrenal tumors, cardiac pheochromocytomas frequently go undiagnosed for years because they exhibit less dramatic clinical features as a result of the absence of phenylethanolamine-N-methyltransferase, which converts norepinephrine into epinephrine. Patients with cardiac pheochromocytoma often undergo several diagnostic tests, sometimes even exploratory laparotomy, before cardiac localization is considered.

In the clinical setting of a patient being referred for a cardiologic workup, TEE proved to be more useful than other tests, because it consistently gave initial confirmation of a mass typical of pheochromocytoma and allowed localization in relation to adjacent cardiac structures. Detection of possible infiltration of the cardiac walls with TEE can be of additional use for surgical decision making. The general search for a suspected pheochromocytoma often begins with computed tomography of the adrenal glands; if the results are negative, MIBG scintigraphy is performed. MIBG is a catecholamine analogue that uses the amine precursor uptake mechanism and may thus be incorporated into vesicles or neurosecretory granules in the cytoplasm. MIBG scintigraphy is highly sensitive, but it does not allow for tumor delineation in relation to adjacent structures and is reported as negative in 10% to 20% of cases. Radiolabeled octreotide, an analogue of somatostatin, is used, although with less specificity, to demonstrate tumors that have soma-
TABLE 1 Characteristics of Eight Patients With Primary Pheochromocytoma of the Heart

<table>
<thead>
<tr>
<th>Case</th>
<th>Age (yrs)</th>
<th>Tumor Location*</th>
<th>Size (cm)</th>
<th>TTE/TEE</th>
<th>MRI/CT</th>
<th>Octr/MIBG</th>
<th>Angio/Blood Supply</th>
<th>Operative Resection</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>25 F</td>
<td>C</td>
<td>2.6 x 2.5 x 4.0</td>
<td>+/- /-</td>
<td>+/- /-</td>
<td>+/- /-</td>
<td>+/- RCA</td>
<td>4</td>
</tr>
<tr>
<td>2</td>
<td>26 F</td>
<td>A,B</td>
<td>1.7 x 2.3 &amp; 1.5 x 1.7</td>
<td>+/- /+</td>
<td>0/+</td>
<td>0/+</td>
<td>+/- LCA &amp; RCA</td>
<td>4</td>
</tr>
<tr>
<td>3†</td>
<td>28 F</td>
<td>A</td>
<td>4.4 x 5.1 x 4.0</td>
<td>+/- /+</td>
<td>+/- /+</td>
<td>+/- /+</td>
<td>+/- LCA &amp; RCA</td>
<td>4</td>
</tr>
<tr>
<td>4</td>
<td>36 M</td>
<td>A</td>
<td>4.0 x 3.8</td>
<td>O/+  /+</td>
<td>O/+  /+</td>
<td>O/+  /+</td>
<td>+/- LCA &amp; RCA</td>
<td>4</td>
</tr>
<tr>
<td>5</td>
<td>39 F</td>
<td>D</td>
<td>5.0 x 4.9</td>
<td>+/- /-</td>
<td>+/- /-</td>
<td>+/- /-</td>
<td>+/- RCA &amp; BR</td>
<td>N</td>
</tr>
<tr>
<td>6</td>
<td>42 F</td>
<td>B</td>
<td>4.0 x 3.5 x 3.3</td>
<td>0/+  /+</td>
<td>0/+  /+</td>
<td>0/+  /+</td>
<td>0/+ RCA</td>
<td>4</td>
</tr>
<tr>
<td>7</td>
<td>47 F</td>
<td>C</td>
<td>2.5 x 2.5</td>
<td>O/+  /+</td>
<td>O/+  /+</td>
<td>O/+  /+</td>
<td>0/+ RCA</td>
<td>N</td>
</tr>
<tr>
<td>8</td>
<td>57 M</td>
<td>C</td>
<td>1.5 x 2.0</td>
<td>+/- /-</td>
<td>+/- /-</td>
<td>+/- /-</td>
<td>+/- RCA</td>
<td>N</td>
</tr>
</tbody>
</table>

*Letters refer to the labels on Figure 1.
†Case 3 has been reported before.
Angio = angiography; CT = computed tomography; LCA = left coronary artery; MRI = magnetic resonance imaging; Octr = octreotide scintigraphy; RCA = right coronary artery; TTE = transthoracic echocardiography; + = tumor detected; — = tumor not detected; 0 = not performed or no information available.

Echocardiography, particularly TEE, disclosed a characteristic appearance of primary cardiac pheochromocytoma (void, granular tumors within the atrioventricular groove adjacent to the epicardial coronary arteries). These characteristic tumor features should be considered extremely typical of cardiac pheochromocytoma and should prompt necessary anatomic and biochemical confirmation as well as assist in the decision between surgical or medical management.

4. Fitzgerald PJ, Ports TA, Cheitlin MD, Magilligan DJ, Tyrrell JB. Intracardiac tostatin receptors on their surface. Magnetic resonance imaging and computed tomography are very useful for detecting and characterizing cardiac pheochromocytoma. However, we observed that echocardiography was the first screening imaging test.
Balance and Gait in Older Adults With Systemic Hypertension*

Jeffrey M. Hausdorff, PhD, Talia Herman, BPT, Rossitza Baltadjieva, MD, Tanya Gurevich, MD, and Nir Giladi, MD

It has been suggested that age-associated abnormalities in blood pressure (BP) homeostasis may precipitate decreases through transient underperfusion of the brain and that hypertension may exacerbate impaired BP regulation and thus increase the risk of falling.1 According to this hypothesis, a transient impairment of BP regulation and thus increase the risk of stroke, Parkinson’s disease, parkinsonism, or any neurologic disease, use of anti-parkinsonian or antispasmodic medications, orthostatic hypotension, cerebellar dysfunction, significant visual or vestibular disturbances, or significant orthopedic disturbances. Subjects with dementia, a history of psychiatric disease, or subjects who were taking antipsychotic medications were also excluded. In addition, we excluded patients with a history of head trauma or clinically documented cerebrovascular events. All subjects were recruited as “healthy controls” to provide a reference group in an ongoing study of neurologic disease in older adults. For the purposes of the present study, these “healthy controls” were stratified into 2 groups. Subjects were classified as “hypertensives” if they were under a doctor’s order to take a prescription medication to control their hypertension (i.e., they reported a history of hypertension and a prescription for BP medications). Other subjects were defined as “normotensives.” Informed written consent was obtained.

To characterize the study population, we obtained a detailed medical history, reviewed all medications, and performed a full, structured neurologic examination including the motor portion (part III) of the Unified Parkinson’s Disease Rating Scale (UPDRS).3 The Charlson Comorbidity Index was determined.4 Mental health was assessed using the Mini Mental State Exam5 and the long form of the Geriatric Depression Scale.6 Isometric muscle strength was assessed at the quadriceps, averaging left and right legs, as previously described.7 The ability to perform activities of daily living was assessed with the timed “get up and go” test; subjects were instructed to get up from a sitting position, walk 3 meters, turn around, and return to sit. The time taken to complete this task was recorded to the nearest 0.1 second. The 10 meter walk test was also performed to evaluate gait and balance.

Subjects were included if they were living in a community, were aged 65 to 90 years, were able to follow instructions, and reported no disturbances in their walking abilities. Exclusion criteria included: history of stroke, Parkinson’s disease, parkinsonism, or any neurologic disease, use of anti-parkinsonian or antispasmodic medications, orthostatic hypotension, cerebellar dysfunction, significant visual or vestibular disturbances, or significant orthopedic disturbances. Subjects with dementia, a history of psychiatric disease, or subjects who were taking antipsychotic medications were also excluded. In addition, we excluded patients with a history of head trauma or clinically documented cerebrovascular events. All subjects were recruited as “healthy controls” to provide a reference group in an ongoing study of neurologic disease in older adults. For the purposes of the present study, these “healthy controls” were stratified into 2 groups. Subjects were classified as “hypertensives” if they were under a doctor’s order to take a prescription medication to control their hypertension (i.e., they reported a history of hypertension and a prescription for BP medications). Other subjects were defined as “normotensives.” Informed written consent was obtained.

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From the Movement Disorders Unit, Department of Neurology, Tel-Aviv Sourasky Medical Center, Sackler School of Medicine, Tel-Aviv University, Tel-Aviv, Israel; Gerontology Division and Margaret and H.A. Rey Laboratory for Nonlinear Dynamics in Medicine and Biology, Beth Israel Deaconess Medical Center and Division on Aging, Harvard Medical School, Boston, Massachusetts. This work was supported in part by Grants AG-14100, RR-13622, HD-39838, and AG-08812 from the National Institutes of Health, Bethesda, Maryland. Dr. Hausdorff’s address is: Movement Disorders Unit, Tel-Aviv Sourasky Medical Center, 6 Weizmann Street, Tel-Aviv 64239, Israel. Email: jhausdorff@bolum.health.harvard.edu. Manuscript received June 27, 2002; revised manuscript received and accepted October 17, 2002.

*Time series used to determine stride and swing time dynamics (e.g., coefficient of variation and fractal indexes) are available for downloading and additional study via an NIH sponsored website: www.physionet.org.