CASE REPORTS

Left Atrial Appendage Aneurysm
Correlation of Noninvasive with Clinical and Surgical Findings:
Report of a Case

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SUMMARY
Congenital left atrial appendage aneurysm is rarely diagnosed on the basis of an abnormal cardiac silhouette. Patients with a left atrial appendage aneurysm often present with symptoms of systemic emboli or supraventricular arrhythmias. A patient with left atrial appendage aneurysm was diagnosed by correlation of two noninvasive techniques, echocardiography and radionuclide scintiscanning. Angiography was confirmatory and aneurysmectomy was successfully performed.

CONGENITAL LEFT ATRIAL ANEURYSMS are rare and may involve the left atrial wall \textsuperscript{6-10} or the left atrial appendage.\textsuperscript{11-13} This discussion is limited to left atrial appendage aneurysms. These aneurysms mimic mediastinal or cardiac tumors. Systemic emboli and supraventricular arrhythmias endanger the patient with this condition.

Recently we had the opportunity to correlate noninvasive diagnostic techniques with clinical and surgical findings in a child with a left atrial appendage aneurysm. Echocardiography in conjunction with radionuclide scintiscanning established the diagnosis which was later confirmed by angiography and surgery.

Case Report
A chest X-ray obtained during evaluation for a moderately severe upper respiratory tract infection in this five-year-old girl revealed a prominent convexity on the upper left heart border. Following resolution of her infection, she was referred to the University of Nebraska Medical Center. The only abnormal physical finding in this normally developed child was a grade I/VI systolic ejection murmur audible only in the supine position at the third left intercostal space. Routine laboratory work and screening tests were within normal limits. The chest X-ray (fig. 1) showed a marked prominence at the upper left heart border. The electrocardiogram (fig. 2) recorded a heart rate of 140 beats/minute, a P-R interval of 0.16 seconds, broad notched "P" waves in lead I and biventricular, broad "P" wave in lead V\textsubscript{a}.

Noninvasive Studies

Echocardiography
The routine echocardiographic scan was normal. The aorta measured 1.9 cm in diameter and the left atrium was 2.0 cm. Left ventricular walls and cavity dimensions were within normal limits. All cardiac valves were normal and there was no evidence of solid tumor masses. The abnormal chest X-ray prompted us to reposition the transducer in the third intercostal space and direct the sound beam lateral to the aorta. The pulmonary artery with the pulmonic valve was seen and was normal in diameter (1.9 cm) (fig. 3). Further lateral rotation showed a large fluid filled cavity measuring 4.3 cm in diameter.

Radionuclide Cardiac Blood Pool Evaluation
Six millicuries of technetium sulphur colloid were injected intravenously and sequential scintigrams were taken in the anterior projection using a gamma camera system. Injections of Diethylenetriaminepentaacetic acid (DTPA) were then accomplished and sequential images were obtained in the lateral and right anterior oblique projections. A large, slow emptying cavity of cardiovascular origin was seen anterior and left of the mediastinum (fig. 4). The radionuclide scintigrams established the roentgenographic abnormality was of cardiovascular origin. The echocar-
diagram established that it was very large (4.3 cm in diameter anteroposteriorly) and was not a pulmonary artery aneurysm.

Consideration of both these studies established that the roentgenographic abnormality was a left atrial appendage aneurysm.

Invasive Studies

Cardiac catheterization yielded no evidence of intracardiac shunts or valvar gradients. The mean pulmonary artery wedge pressure was 14 mm/Hg (table 1). Biplane pulmonary artery cine angiography with levophase study showed filling of a massive left atrial appendage aneurysm which compressed and displaced the left ventricle (fig. 5). Left ventricular

<table>
<thead>
<tr>
<th>Catheter position</th>
<th>Pressure (mm Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>RA “a” wave</td>
<td>8, mean 4</td>
</tr>
<tr>
<td>RV</td>
<td>37/0–7</td>
</tr>
<tr>
<td>PA</td>
<td>32/13, mean 20</td>
</tr>
<tr>
<td>LV</td>
<td>135/0–12</td>
</tr>
<tr>
<td>Ao</td>
<td>135/85</td>
</tr>
<tr>
<td>PAW “a” wave</td>
<td>16</td>
</tr>
<tr>
<td>“v” wave</td>
<td>20, mean 14</td>
</tr>
</tbody>
</table>

Abbreviations: RA = right atrium; PAW = pulmonary artery wedge; RV = right ventricle; PA = pulmonary artery; LV = left ventricle; Ao = aorta.

Table 1

Hemodynamic Data

Figure 1

Top) The preoperative PA and lateral chest X-rays revealed prominent convexity at the upper left heart border. Bottom) The postoperative chest X-rays showed decreased prominence at the upper left heart border.

Figure 2

Top) Preoperative electrocardiogram recorded a heart rate of 140 beats/min, P-R interval of 0.16 sec and broad notched “P” waves in lead I. Leads V1–V6 are taken at ½ standardization (i.e., 1 mV = 0.5 cm). All other leads are at full standardization (1 mV = 1 cm). An example of full standardization is shown in lead II. Bottom) Postoperative electrocardiogram recorded a heart rate of 125 beats/minute, P-R interval of 0.13 sec and the absence of “P” wave notching. Leads V2, V4, and V6 are taken at ½ standardization. All other leads are taken at full standardization. An example of full standardization is shown in lead II.
angiography confirmed compression and displacement of the ventricle. Mitral valve abnormalities were absent.

Surgical exposure through a median sternotomy found the pericardium intact. A large, thin-walled left atrial appendage aneurysm measuring 7 cm × 7 cm × 4 cm was found, compressing the left ventricle and forcing it posteriorly and medially (fig. 6). Under cardiopulmonary bypass the aneurysm was excised at a "neck" which joined the aneurysm to the body of the left atrium. Inspection revealed no evidence of thrombus formation.

Pathological examination revealed a 0.4 mm thick aneurysm wall joining to a left atrial wall which was 2 mm thick. Microscopic examination of the aneurysm showed areas of normally appearing atrial muscle interspaced with areas of fibrous tissue with complete absence of myocardial fibers.

The postoperative course was uneventful. The systolic murmur persisted. The chest X-ray (fig. 1)
showed decreased prominence at the upper left heart border. The postoperative electrocardiogram (fig. 2) recorded a heart rate of 125 beats/minute, a P-R interval of 0.13 seconds, and no "P" wave notching. There was an increase in the lateral ventricular voltage. The postoperative routine echocardiogram remained within normal limits. The echocardiographic technique used preoperatively was employed and there was no cavity demonstrated lateral to the pulmonary artery.

Discussion

An abnormal roentgenographic cardiac silhouette should always concern the clinician. Dilatation of the left atrium causes an abnormal cardiac silhouette. Left atrial enlargement commonly occurs as a sequela to mitral valve disease or left ventricular myocardial disease; rarely, the enlargement is a congenital left atrial aneurysm. These aneurysms may involve either the left atrial wall or the left atrial appendage.8-16

Review of the 12 reported patients,8-16 including our patient, with left atrial appendage aneurysm revealed a propensity for embolic events and supraventricular arrhythmias (table 2). These potentially lethal complications call for prompt diagnosis and treatment. Atrial thrombi occur in patients with atrial arrhythmias. Even in the absence of arrhythmias emboli have occurred in patients with left atrial appendage aneurysms. Stasis of blood within the aneurysm probably predispose to thrombus formation.

The systolic murmurs which were audible in most patients with left atrial appendage aneurysms were probably secondary to causes other than the aneurysm. Cine angiographic analysis shows slow flow
### Table 2

**Clinical, Angiographic, and Surgical Findings in Patients with Left Atrial Appendage Aneurysm**

<table>
<thead>
<tr>
<th>Author &amp; date</th>
<th>Age/ Sex</th>
<th>SVA</th>
<th>Presentation</th>
<th>Physical findings</th>
<th>ECG</th>
<th>Chest X-ray</th>
<th>Clinical diagnosis</th>
<th>Angio</th>
<th>Preop. diagnosis</th>
<th>Surgical finding</th>
<th>Procedure</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Palacio et al.</td>
<td>32</td>
<td>AF</td>
<td>Cerebral emboli &amp; AF</td>
<td>SM in PA area &amp; apex</td>
<td>AF</td>
<td>Typical</td>
<td>None stated</td>
<td>LAA</td>
<td>LAA; clot present</td>
<td>Excision</td>
<td>Reversion to sinus rhythm w/ quinidine</td>
<td>Good result NSR</td>
</tr>
<tr>
<td>Parmley</td>
<td>9</td>
<td>M</td>
<td>Near syncope; emboli p</td>
<td>3/6 SM at apex, ASD</td>
<td>IVCD</td>
<td>Globular, cardiomegaly</td>
<td>ASD &amp; enlarged LA</td>
<td>ASD</td>
<td>ASD &amp; LAAA</td>
<td>Excision &amp; repair ASD</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Williams</td>
<td>27</td>
<td>M</td>
<td>Routine chest X-ray</td>
<td>2/6 SM in PA area</td>
<td>Normal</td>
<td>Typical</td>
<td>PA dil.</td>
<td>LAAA</td>
<td>No surgery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Williams</td>
<td></td>
<td></td>
<td>a. routine X-ray &amp;</td>
<td></td>
<td>Normal</td>
<td>Typical</td>
<td>Cardiac tumor</td>
<td>Cardiac tumor</td>
<td>a. LAAA 7x5 cm</td>
<td>a. None</td>
<td>a. SVT &amp; cerebral emboli 2 yr. later</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>cerebral emboli 15 yr.</td>
<td></td>
<td>Normal</td>
<td>No change</td>
<td>No change</td>
<td>Same</td>
<td>Same</td>
<td>b. Fibrosis of appendage; clots present</td>
<td>b. Excision</td>
<td>b. No additional emboli</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>earlier</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>b. LAAA</td>
<td>Excision</td>
<td>Uneventful recovery</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Chest pain &amp; palp.</td>
<td></td>
<td>Normal</td>
<td>Typical</td>
<td>Mitral valve disease</td>
<td>LAAA</td>
<td>LAAA; peri defect</td>
<td>LAAA</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Godwin, et al.</td>
<td>20</td>
<td>F</td>
<td>Tachycardia &amp; assoc.</td>
<td></td>
<td>Normal</td>
<td>Typical Lat. decubitus view surg. extra cardiac mass</td>
<td>None stated</td>
<td>LAAA</td>
<td>LAAA 8x5 cm</td>
<td>Excision</td>
<td>No palpitation</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>chest pain Palp.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>No surgery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sloman, et al.</td>
<td>23</td>
<td>M</td>
<td>Routine X-ray</td>
<td>SM at base</td>
<td>PVC's &amp; &quot;T&quot; wave abnormality</td>
<td>Typical</td>
<td>PAA or LAAA</td>
<td>LAAA</td>
<td>No surgery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hall</td>
<td>22</td>
<td>M</td>
<td>Cerebral emboli</td>
<td>No cardiac abnormalities</td>
<td>Normal</td>
<td>Typical</td>
<td>None stated</td>
<td>Lesion of LV</td>
<td>Lesion of LV</td>
<td>LAA; no clots</td>
<td>Excision</td>
<td></td>
</tr>
<tr>
<td>Salonikides, et al.</td>
<td>34</td>
<td>F</td>
<td>PAT</td>
<td>Normal</td>
<td>Normal</td>
<td>Typical</td>
<td>Cardiac tumor. Aneur of LAA, PA, LV, or AO</td>
<td>LAAA</td>
<td>No surgery</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Scanderud, et al.</td>
<td>43</td>
<td>F</td>
<td>AF, DOE, ankle edema</td>
<td>2/6 SM LSB</td>
<td>AF</td>
<td>Cardiomegaly</td>
<td>Cardiomegaly</td>
<td>Mass opacified during left heart filling</td>
<td>LAAA</td>
<td>Excision</td>
<td>5 wks. post-op reverted to NRS</td>
<td></td>
</tr>
</tbody>
</table>

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congenital left atrial appendage aneurysm. It would be unlikely that a Reynolds number sufficient to cause a murmur could be reached.

The chest X-ray in all patients showed a prominent convexity on the left heart border or globular cardiomegaly.

The electrocardiograms have usually been normal or revealed supraventricular arrhythmias. Two patients had interventricular conduction defects. Our patient is the first to show evidence of left atrial abnormalities. The increase in lateral ventricular forces in our patient is apparently a result of the return of the left ventricle to its normal position following aneurysmectomy.

Echocardiography has proven valuable in the evaluation of mitral valve disease, and in patients with intracardiac tumors such as left atrial myxoma, right atrial myxoma, and ventricular rhabdomyoma. Recently, the echocardiographic findings in patients with extracardiac mediastinal masses such as thymic cysts, fibrolipoma, or fibrolipoma, have been described. Although not reported, the echocardiographic diagnosis of pulmonary artery aneurysm should be possible. The echocardiogram was extremely valuable in the evaluation of our patient with a left atrial appendage aneurysm. The noninvasive diagnosis requires correlation of the echocardiogram with the radionuclide studies. In our patient pericardial and extracardiac cysts were diagnostic possibilities on the basis of the echocardiogram alone but were excluded by the radionuclide studies. Conversely, pulmonary artery aneurysm was a possible diagnosis from the radionuclide studies only but was excluded by echocardiography. Thus the diagnosis of left atrial appendage aneurysm was established. Cardiac catheterization was confirmatory.

Surgical excision is recommended to eliminate a potential source of emboli and for relief of supraventricular arrhythmia. Surgery is an effective therapeutic measure and there are no reported operative mortalities. Aneurysmectomy was postponed in one reported patient, and subsequently he suffered a cerebral embolus.

Partial left pericardial defect with left atrial appendage herniation may clinically and radiographically simulate left atrial appendage aneurysm. The herniated appendage rarely reaches the size of the left atrial appendage aneurysm. Therefore, echocardiography would provide a means of establishing the diagnosis.

Conclusion

Congenital left atrial appendage aneurysm is a rare cause of an abnormal roentgenographic cardiac silhouette. Patients with this entity may suffer from
systemic emboli and supraventricular arrhythmias. Correlation of the echocardiographic and radionuclide findings can establish the diagnosis of left atrial appendage aneurysm. We present a patient with left atrial appendage aneurysm which was diagnosed by noninvasive techniques, with confirmation by angiography and at surgery. The aneurysm was successfully excised.

Acknowledgment

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