Large Anomalous Fibrous Sac in the Right Side of the Heart

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SUMMARY
In an adult with atrial septal defect, an unusual conical-shaped fibrous sac resembling a closed windsock occupied the right ventricle and main pulmonary artery. The base of the sac being attached by fibrous strands to the right atrium suggests that the structure is derived from the right valve of the sinus venosus.

The sac is considered to have filled with blood, thereby reducing potential right ventricular diastolic volume.

It is probable that the sac emptied into the right atrium during right ventricular systole, thereby creating a functional state comparable to that of tricuspid insufficiency. Occupation of right ventricular space by the sac established a state somewhat comparable to that in Ebstein's anomaly of the tricuspid valve.

Additional Indexing Words:
Chiari's network  Atrial septal defect  Sinus venosus
Inflow obstruction, right ventricle  Ebstein's anomaly of tricuspid valve

dyspnea with exercise of 9 months' duration. Mild dyspnea had been present since the age of 3 years, but no thoracic pain or episodes of unconsciousness had occurred. Physical examination revealed the brachial blood pressure to be 144 mm Hg, systolic and 94 mm Hg, diastolic.

The pulse rate was 80 beats per min. A soft systolic murmur was present at each sternal border, radiating to the right side.

The electrocardiogram showed incomplete right bundle-branch block and right ventricular hypertrophy.

Following injection of radiopaque material into the right atrium and into the main pulmonary artery, there was opacification of the left atrium and there was poor visualization of the pulmonary artery. The findings were interpreted as indicative of atrial septal defect with right ventricular overload and possibly outflow obstruction to the right ventricle.

The latter, however, was not confirmed by studies of pressure, which showed moderately elevated values both in the right ventricle and pulmonary artery (table 1). Studies for levels of oxygen saturation were indicative of a left-to-right shunt at the atrial level. The levels of oxygen saturation in the upper and lower superior vena cava were 61.5 and 63.2%, respectively. In the right atrium, right ventricle, and pulmonary artery, the values were 79.7, 79.7, and 82.4% in that order. Blood in a systemic artery showed a value of 95.8% oxygen saturation.

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Table 1

Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressures (mm Hg)</th>
<th>Systolic</th>
<th>Diastolic</th>
<th>Mean</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary wedge</td>
<td>18</td>
<td>6</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>44</td>
<td>20</td>
<td>28</td>
<td></td>
</tr>
<tr>
<td>Right ventricular infundibulum</td>
<td>44</td>
<td>0-4</td>
<td>25</td>
<td></td>
</tr>
<tr>
<td>Right ventricle, low</td>
<td>52</td>
<td>5</td>
<td>26</td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>10</td>
<td>0</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>Right femoral artery</td>
<td>160</td>
<td>90</td>
<td>115</td>
<td></td>
</tr>
</tbody>
</table>

Using cardiopulmonary bypass, the atrial septal defect, which was of the fossa ovalis type, was closed. A weblike structure was noted adjacent to the defect and passed out of the field of vision toward both the superior vena cava and the pulmonary artery. Cardiac arrest occurred on the day following operation, and resuscitative efforts were unsuccessful.

Pathologic Features

Except for pulmonary congestion and edema, the pertinent findings were confined to the heart.

The great veins were normally connected to the right atrium, and the wall of the right atrium was considerably thickened. A circular patch of synthetic material had been sutured to the right side of the atrial septum in order to cover a 2 cm diameter atrial septal defect at the fossa ovalis. The valve of the foramen ovale was short, accounting for the principal defect. In addition, several smaller perforations were present in the valve of the foramen ovale.

The unusual feature of the specimen was the presence of a large white, fibrous, cone-shaped, windsock-like sac of which the distal end was closed. The sac occupied the right ventricle and the proximal part of the pulmonary artery, the broader and open base lying proximally (figs. 1 and 2). The sac measured about 6.5 cm in length. The diameter of its inlet was about 4.5 cm, and the width of the distal end, lying in the pulmonary artery, was about 3.0 cm. The capacity of the sac was about 30 ml. Its wall measured from 1 to 2 mm in thickness. Three fibrous bands, after passing through the orifice of the tricuspid valve, connected the base of the sac with the right atrial wall (figs. 1a and 2).

The first two of these bands began as several discrete strands resembling chordae. These fused to form a solitary cord and then divided to form the two separate bands. The longer of these bands was 3 cm long and inserted at the crista terminalis. The shorter of these bands was 1.5 cm long and inserted on the anterior aspect of the atrial septum 1 cm above the tricuspid valve. The third band inserted in the atrial septum between the atrial septal defect and the septal leaflet of the tricuspid valve. In addition to the above bands, two larger strands emanated from the inferior portion of the free margin of the proximal end of the sac. These were found transected and the exact point of natural attachment could not be determined. It is likely, however, that these were attached to the atrial wall near the coronary sinus and were transected at the time of operation.

The sac followed the course of blood flow through the right side of the heart; in other words, through the right ventricle, the infundibulum, the pulmonary valve, and into the pulmonary trunk for a distance of about 2 cm (fig. 1a).

The strands running from the right atrium to the base of the sac crossed the tricuspid valve at the junction of the posterior and septal leaflets. In this region, the valvular tissue was thickened and partially adherent to the related strands.

The sac was partially adherent in linear fashion to the endocardium along the posterior wall of the infundibulum and to the lining of the pulmonary trunk. At the pulmonary valve, the attachment coursed between the right and left cusps of the valve. The terminal portion of the sac was found to be closed.

The cusps of the pulmonary valve were opaque on the basis of mild and uniform fibrous thickening. No fusion between the cusps was present. The mitral and aortic valves were normal.

The heart was hypertrophied, weighing 525 g. The enlargement resulted mainly from the nature of the right ventricle of which the wall was rather uniformly hypertrophied, measuring 1.0 cm in thickness. The left ventricular wall measured 1.6 cm in thickness.

The cavity of the right ventricle was moderately enlarged. The right atrial chamber was only slightly enlarged, failing to show the degrees of enlargement commonly observed in classical examples of atrial septal defect.

Comment

In 1897 Chiari described, in detail, the fibrous network in the right atrium which now carries his name. The Chiari network may be defined as a network of fine or coarse fibers in the right atrium with attachments extending from the region of the crista terminalis to the valves of the inferior vena cava and coronary sinus, or even to the floor of the right atrium.
anomalous fibrous sac

Figure 1

a. Interior of right ventricle (R.V.) and pulmonary trunk (P.T.). A large fibrous sac (S.) passes through the tricuspid valve (T.V.), the right ventricle, and into the pulmonary trunk. Distal end of sac has been opened. b. Interior of right atrium (R.A.) and right ventricle (R.V.). Base of sac (S.) lies at level of tricuspid valve (T.V.) and is attached to right atrial wall by fibrous strands. Apex of sac disappears toward pulmonary valve. Probe in coronary sinus.

In the region of the orifice of the coronary sinus (fig. 3).

Developmentally, the right valve of the sinus venosus evolves into the valve of the inferior vena cava and the valve of the coronary sinus, a process which involves considerable reduction in size of the valve. In this changing process the tissue undergoes a fenestrating process so that a network (Chiari's net) may be formed from remnants that usually disappear.

In the case presented, it is assumed that in the transformation of the right valve of the sinus venosus minimal resorption or fenestration occurred, leaving the net an imperforate sheet which could be stretched forward by the current of blood, as a spinnaker is blown forward by the wind, according to Jones and Niles.

In our case, attachment of the structure to the infundibular area and to the pulmonary artery is considered a reaction to trauma by the sac moving against the lining of the involved areas.

In addition to the case of Jones and Niles, we are aware of only one other reported instance of a giant remnant of the right valve of the sinus venosus. This is the case of Goforth who, in 1926, described a free fibrous cord originating in the right atrium, passing through a patent foramen ovale, the left atrium and left ventricle, and terminating in the aorta. Although the etiology of this
anomaly was not determined, it was probably a remnant of the Chiari network.

It is of interest to consider the possible functional effects of the peculiar sac in our case.

One may assume that the sac filled with blood during ventricular diastole, thereby occupying right ventricular space. In this way it may have been responsible for less forward flow than was the potential for the right ventricle, a feature comparable to that in Ebstein’s anomaly of the tricuspid valve. The poor opacification of the pulmonary artery after injection of contrast material into the right atrium is in keeping with such a function.

During systole two possibilities must be entertained. The first was that the tricuspid valve closed around the base of the sac, thereby trapping blood in the sac. In this way the sac could have been responsible for pulmonary stenosis. This fact is denied by the data of the cardiac catheterization.

The second possibility is that blood in the sac regurgitated into the right atrium. The latter would have had an effect upon the right ventricle comparable to that of tricuspid insufficiency.

References
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