Case Report

The Role of Transesophageal Echocardiography in Diagnosis and Treatment in a Series of Patients with Chronic Thromboembolic Pulmonary Hypertension

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Abstract

We describe 6 cases of chronic thromboembolic pulmonary hypertension, who underwent pulmonary thromboendarterectomy in our center. Transthoracic and transesophageal echocardiography provided valuable data on surgical accessibility of thrombus, its extension, chronicity and hemodynamic consequences on right ventricular function. Transesophageal echocardiography as a rapid, bedside and easily available method has been a useful guide for diagnosis and guiding the treatment for these patients.

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Introduction

Chronic thromboembolic pulmonary hypertension (CTEPH) results from obstruction of the pulmonary vascular bed by nonresolving thromboemboli which represents a potentially correctable cause of pulmonary hypertension. CTEPH occurs much more frequently after acute pulmonary embolism that had been classically believed. New data indicate the frequency of CTEPH between 1 and 4 percent. However, the estimated number of unreported cases may be as high. Right heart failure and death are consequences of CTEPH if patients are not treated. Medical therapy for pulmonary hypertension caused by pulmonary vascular occlusion is generally unsatisfactory and palliative. In contrast to idiopathic pulmonary arterial hypertension (iPAH), which manifests in pulmonary vessels of <300 µm diameter, CTEPH has been initially discriminated from PAH by its major-vessel involvement of the vascular remodeling process, rendering it accessible to surgical intervention with removal of the obstructing lesions. Therapy by pulmonary endarterectomy offers a surgical cure, yet this procedure is not commonly applied. It has been appreciated for many years that CTEPH may not be explained simply by pulmonary vascular obliteration due to unresolved thrombemboli. CTEPH has emerged as a ‘dual’ pulmonary vascular disorder with major vessel vascular remodeling of thrombus organization, combined with a small vessel pulmonary arteriopathy that is a target for classic vasodilator treatments. A precise evaluation of medical history and a thorough physical examination are keys for making a diagnosis. Transthoracic echocardiography is an important diagnostic tool for pulmonary hypertension, and is also a good screening method. It is usually the first study to suggest that an abnormality of the pulmonary vasculature is present. Depending on the stage of the disease when echocardiography is performed, it may demonstrate variable degrees of right atrial and right ventricular enlargement, abnormal right ventricular systolic function, tricuspid regurgitation, a leftward displacement of the interventricular septum, decreased left ventricular size, and abnormal left ventricular systolic and diastolic function. Transesophageal echocardiography holds the promise of diagnosing pulmonary embolism by direct visualization of a thrombus rather than by relying on indirect signs, such as right ventricular enlargement and hypokinesis. The examination assesses the extent of thromboembolism as well as its surgical accessibility. TEE has a definite role in the management of patients with acute pulmonary thromboembolism or in pulmonary embolism associated with right-sided intracardiac masses and in the selection of patients with PH for pulmonary thromboendarterectomy.

Case Series

Here we describe six cases of CTEPH admitted to our heart center. Diagnosis was made based on history, physical examination, chest x-ray, transthoracic and transesophageal echocardiography. All patients presented with progressive dyspnea on exertion and signs of right side heart failure including fatigue, palpitations, syncope, or edema. There had been history of pulmonary thromboembolism in all of them and there have been treated with warfarin for at least three months but represented with progressive dyspnea and persistent pulmonary hypertension. After detection of large obliterating thrombus in PA and its branches with PAH, patients were assessed for possibility of surgical intervention. Patient selection for pulmonary thromboembolectomy was accomplished according to the following criteria: 1- evidence of surgically accessible thrombi (by TEE); 2- absence of severe comorbidities; 3- symptomatic severe pulmonary hypertension; 4- history of pulmonary thromboembolism and treatment with anticoagulant for at least 3 months; 5- patient informed consent

Patients’ characteristic data are shown in Table. Their ages range from 32 to 72. They all had
history of pulmonary thromboembolism and had been treated with oral anticoagulants for at least 3 months. They presented with progressive dyspnea in functional class of III (3 patients) or IV (3 patients) in a relatively unstable condition. They all underwent TTE & TEE pre-operation, the diagnosis was made based on echocardiographic findings. There was severe pulmonary hypertension with severe right ventricular dysfunction present in all cases. Predisposing factor of protein C deficiency was identified in one of the cases.

Patients Characteristics:

<table>
<thead>
<tr>
<th>Case</th>
<th>Gender</th>
<th>Age</th>
<th>PAP (pre-op)</th>
<th>PAP (post-op)</th>
<th>IVC filter /plication</th>
<th>Associated conditions</th>
<th>Operation complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Male</td>
<td>53</td>
<td>120</td>
<td>27</td>
<td>-</td>
<td>Previous CVA, Large PFO</td>
<td>TIA</td>
</tr>
<tr>
<td>2</td>
<td>Male</td>
<td>39</td>
<td>100</td>
<td>50</td>
<td>plication</td>
<td>Protein C deficiency</td>
<td>-</td>
</tr>
<tr>
<td>3</td>
<td>Male</td>
<td>33</td>
<td>110</td>
<td>75</td>
<td>-</td>
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<td>-</td>
</tr>
<tr>
<td>4</td>
<td>Male</td>
<td>72</td>
<td>100</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>expired soon after surgery</td>
</tr>
<tr>
<td>5</td>
<td>Female</td>
<td>46</td>
<td>100</td>
<td>40</td>
<td>filter</td>
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<td>-</td>
</tr>
<tr>
<td>6</td>
<td>Male</td>
<td>32</td>
<td>65</td>
<td>45</td>
<td>plication</td>
<td>PFO</td>
<td>-</td>
</tr>
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</table>

All patients underwent pulmonary thromboendarterectomy and IVC filter was implanted in 1 patient as it was not available in the remaining cases. Inferior vena cava plication was performed in 2 cases as an alternative approach. The procedure was performed through median sternotomy, cardiopulmonary bypass, deep hypothermia induction and the right and left thromboendarterectomies were performed sequentially under complete circulatory arrest. The obstructing material (the organized thrombus) was extracted. Distal circumferential dissection was performed for complete removal, and afterwards the arteriotomy site was closed with pericardial patch to restore the normal circumference of the vessel. After surgery patients were transferred to cardiac surgery intensive care unit for post operative care. One of our patients died shortly after surgery of cardiogenic shock and one with a history of ischemic stroke, experienced transient ischemic attack. Others had rather uncomplicated hospital stay and were discharged in good condition and in post-op follow up they experienced significant improvement in functional capacity and exercise tolerance. Post operative TEE was performed and showed improved RV function in all surviving cases.

Discussion

Chronic thromboembolic pulmonary hypertension (CTEPH) is characterized by intraluminal thrombus organization and fibrous obliteration of pulmonary arteries. Although medical therapy for the condition is supportive only, surgical therapy is curative. No prospective studies have evaluated the most appropriate diagnostic approach to CTEPH, but a diagnostic algorithm has been proposed in the meeting of a group of international experts in Zurich, in 2005. The algorithm suggests patients with unexplained pulmonary hypertension or pulmonary hypertension and a history of pulmonary thromboembolism should undergo ventilation-perfusion scintigraphy. There is consensus among experts that a normal ventilation-perfusion scintigram practically rules out the presence of CTEPH. In contrast, the presence of multiple bilateral perfusion defects makes CTEPH the most likely diagnosis. If scintigraphy shows indeterminate results, the next diagnostic step is usually CT angiography. Pulmonary angiography remains a standard diagnostic tool in the assessment of patients with probable or definite CTEPH both to establish the diagnosis and to assess operability which should be performed by experienced staff. Although pulmonary angiography is considered the standard diagnostic tool in CTEPH, it’s not risk-free.
and should be performed by experienced staff. Considering the lack of a referral center in our vicinity for CTEPH patients, referring to a specialized referral center were almost out of question. Considering the lack of enough experience in management of these patients, and the reluctance to perform pulmonary angiography we used TEE as a guide to help diagnosis and management of these patients. TEE as a non-invasive, time-saving and bedside method provided valuable information on anatomic details of thrombus, its extension, texture, chronicity, hemodynamic consequences and surgically availability.

References