Pulmonary Artery Thrombi Detected by Echocardiography in Patients with Pulmonary Hypertension Secondary to Atrial Septal Defect

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This report presents three patients with severe pulmonary hypertension secondary to atrial septal defect associated with thrombus and spontaneous echo contrast within the pulmonary artery diagnosed by transthoracic and transoesophageal echocardiography. Clinical and echocardiographic features seem to suggest local thrombus formation within the pulmonary arteries as a direct consequence of pulmonary hypertension rather than venous thromboembolism.

Key Words: pulmonary artery; thrombus; atrial septal defect; pulmonary hypertension; echocardiography.

Introduction

Thrombi within the pulmonary artery diagnosed by echocardiography have been reported in patients with pulmonary thromboembolism, primary pulmonary hypertension and chronic obstructive pulmonary disease.[1–8] The majority of pulmonary artery thrombi detected by echocardiography have been reported to be secondary to pulmonary embolism.[1–6] Pulmonary embolism and deep venous thrombi are different manifestations of a common disease, namely venous thromboembolism.[7,8] Large clinical studies revealed that echocardiographic data were available in 47–74% of the patients with pulmonary embolism,[1–6,9–16] echocardiographic criteria may distinguish acute from subacute pulmonary embolism, and may predict long-term outcome.[8–16] However, thrombus formation within the pulmonary artery associated with pulmonary hypertension secondary to intracardiac shunt has not been previously reported.

We describe three patients with severe pulmonary hypertension secondary to atrial septal defect associated with thrombus formation within the pulmonary artery as assessed by transthoracic and transoesophageal echocardiography.

Methods

Three patients with long-standing cyanosis, dyspnoea, and findings of severe pulmonary hypertension and right heart failure had been referred to the echocardiography laboratory. Erythrocytosis and systemic arterial blood desaturation were detected in all of the patients. None of the patients had a history of acute deterioration or clinical presentation compatible with acute massive pulmonary embolism. Electrocardiograms revealed right ventricular hypertrophy with systolic strain, clockwise rotation, and right axis deviation. Chest roentgenograms showed enlarged main pulmonary trunk, right and left pulmonary arteries. The transthoracic and transoesophageal echocardiograms showed a prominent left pulmonary artery with a 3 cm diameter. Moderate tricuspid regurgitation was detected in all the patients. Left atrial enlargement and patent foramen ovale were also noted.

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Transoesophageal echocardiography was performed using 3.25 MHz transthoracic and 5 MHz multiplane transoesophageal echocardiography transducers connected to a Vingmed CFM 800 (Horten, Norway) system. Thrombi were diagnosed by the presence of well circumscribed, uniformly consistent, echo-reflective mass with texture distinct from the pulmonary artery wall. Size of the atrial septal defect, flow direction and velocities through septal defect were determined by multiplane transoesophageal echocardiography. Perfusion lung scintigraphy was performed by 99m-Tc Technetium-labelled macroaggregates of albumin, and no perfusion defect which may suggest pulmonary embolism, was documented. Doppler was applied to identify the lower limb veins, but B-mode compression ultrasound study was principally performed. In B-mode compression ultrasound studies the femoral and popliteal veins were found to be compressible, and no hyper-echogenic signal inside the lumen, compatible with thrombus, was detected. A thrombus in the superior and inferior vena cava, in the right atrium, and in the right atrium appendage or right ventricle was excluded by transthoracic and transoesophageal echocardiography. Combined heart and lung transplantation could not be performed.

Case 1
A 61-year-old male was referred with chronic dyspnoea, cyanosis, and signs of right-sided heart failure in November 1995. Acute deterioration in symptoms and history of pleuritic chest pain were not documented. Transthoracic echocardiography showed ostium secundum type atrial septal defect, enlarged main, right and left pulmonary arteries, and enlarged right atrium and right ventricle. Estimated pulmonary artery systolic pressure calculated from severe tricuspid regurgitation was 120 mmHg. Left ventricular dimensions and systolic function were normal. Mass compatible with thrombus within the right pulmonary artery was detected by transthoracic echocardiography. Transoesophageal echocardiography revealed spontaneous echo contrast within the right pulmonary artery, and thrombus filling nearly half of the lumen (Fig. 1). Bidirectional flow through the atrial septal defect was also observed. The patient was diagnosed with Eisenmenger syndrome and discharged on warfarin and vasodilator medication. He died in September 1998. Permission for autopsy was not granted.

Case 2
A 28-year-old female had been operated on with the diagnosis of ostium secundum type atrial septal defect (ASD) in another heart centre in mid-1988, and was referred in June 1996 with fatigue, exertional dyspnoea, cyanosis, and signs of chronic right-sided heart failure.

Transthoracic echocardiography showed marked enlargement in the right atrium, in the right ventricle, and in the pulmonary artery, thrombus within the proximal right and left pulmonary arteries, and severe tricuspid regurgitation with a Doppler estimated pulmonary artery systolic pressure of 70 mmHg [Fig. 2(a,b)]. Bi-directional flow through the interatrial septum compatible with patch failure and spontaneous echo contrast within the main pulmonary artery were also seen. Transoesophageal echocardiography confirmed these findings, and demonstrated a thrombus extending distally to the right pulmonary artery and partially occluding the left pulmonary artery. The patient declined reoperation and was discharged on warfarin and vasodilator therapy.

Case 3
A 49-year-old female was hospitalized in August 1994 with easy fatigue, dyspnoea, cyanosis, ascites and longstanding peripheral oedema. Transthoracic echocardiography revealed severe enlargement in the right heart chambers, main pulmonary arteries with its branches, and presence of drop-out on interatrial septum. Severe pulmonary and tricuspid regurgitation were present, and estimated pulmonary artery systolic pressure was 120 mmHg. Transoesophageal echocardiography showed ostium secundum type atrial septal defect with bidirectional shunt flow. Obliterating thrombi and spontaneous echo contrast within the right pulmonary artery were seen (Fig. 3). Flow velocities in the pulmonary veins were found to be decreased and spontaneous echo formation in the left upper pulmonary vein and left atrium was also detected by transoesophageal echocardiography. The patient was diagnosed with Eisenmenger’s syndrome and was discharged with the
same regimen. Follow-up has failed since 1996. All data concerning the clinical and echocardiographic characteristics of patients are summarized in Table 1.

Discussion

We speculate that clinical and echocardiographic features of our cases appear to suggest local thrombus formation within the pulmonary arteries as a possible consequence of Eisenmenger’s physiology rather than embolization of the thrombi formed within the deep venous system, or right heart chambers.

In patients with a typical clinical history, non-compressible deep limb veins in venous ultrasound, and perfusion defects or ventilation / perfusion mismatch in lung scintigraphy, echocardiography may reveal acute pressure overload and thrombi in-transit, and usually confirms the diagnosis of pulmonary embolism[1–6,9–16]. The typical echocardiographic features of haemodynamically significant pulmonary embolism include dilated and hypokinetic right ventricle, an increased right ventricle/left ventricle ratio due to interventricular septal bulging into the left ventricle, dilated proximal pulmonary arteries, increased velocity jet of tricuspid regurgitation (3–3·5 m.s⁻¹), decreased inspiratory collapsibility of the dilated inferior vena cava, and disturbed ejection pattern (acceleration time <60 ms) in the right ventricular outflow tract[9–16]. Hypokinesis of the right ventricular free wall was reported in 90% of patients with haemodynamically significant pulmonary embolism[9–13]. It is suggested that relatively preserved systolic function in the apical segment of right ventricular wall may differ acute pulmonary embolism from other causes of right ventricular pressure overload[13]. Moreover, the use of criteria such as right free wall thickness >5 mm, tricuspid regurgitation jet velocity >3·7 m.s⁻¹, the presence of both a dilated right ventricular cavity with normal interventricular septal motion, or an inspiratory collapsibility of the inferior vena cava, subacute massive pulmonary embolism was reported to be identified in the majority of the patients[9–16]. Echocardiography may also visualize proximal pulmonary artery thrombi or right heart thrombi in-transit from systemic veins to pulmonary arteries[1–6]. In transoesophageal echocardiographic series, because of the imaging difficulty of the left pulmonary artery due to the shielding effect of the left main bronchus, the reported frequency of thrombi located in the right pulmonary artery was found to be higher than that in left pulmonary artery[6,9–12]. It is
reported that, in comparison to spiral computed tomography, transoesophageal echocardiography had lower sensitivity (79% vs 97.5%), but higher specificity (100% vs 90%)[5]. Although transthoracic echocardiography, in our cases, allowed diagnosis of the atrial septal defect and thrombi in pulmonary arteries, multiplane transoesophageal echocardiography produces high resolution images of the main and proximal left pulmonary artery and longer segments of the right pulmonary artery, permitting direct visualization of the thrombi and atrial septal defect with its bidirectional flow characteristics by colour Doppler imaging. In such patients with clinical findings compatible with Eisenmenger’s syndrome, transoesophageal echocardiography, with its superiority in diagnosing thrombi, septal defect and direction of shunt flow, may obviate the risk of periprocedural paradoxical or pulmonary embolism relating to right heart catheterization and pulmonary angiography.

Thrombi within the central pulmonary arteries have also been documented in patients with chronic obstructive pulmonary disease and primary pulmonary hypertension[7,8]. In our patients, presence of spontaneous echo contrast and thrombus within the pulmonary arteries seemed to be associated with flow stagnation within the pulmonary arteries as a consequence of Eisenmenger’s physiology. Presence of the spontaneous echo contrast within the pulmonary artery in all patients, and within the left upper pulmonary vein in one patient, was considered to be associated with impaired flow in the pulmonary artery beyond the level of thrombotic obliteration.

Neither compressible deep veins on lower limb ultrasound nor negative perfusion lung scan completely exclude the possibility of asymptomatic venous thromboembolic disease[9]. Although compression ultrasonography has high sensitivity (95%) and specificity (98%) for deep venous thrombi in symptomatic patients, its sensitivity reported to be lower (30–50%) in suspected pulmonary embolism[9]. Ventilation lung scintigraphy was not performed in these cases. However, the PISAPED trial and other three studies comprised of large series of patients, have specifically confirmed the validity of a normal perfusion lung scan in ruling out pulmonary embolism[17–20]. Chronic thromboembolic pulmonary hypertension is reported to be more common than generally believed, and should be considered in differential diagnosis of pulmonary hypertension even in the presence of compressible veins[9,15,16]. However, increased pulmonary artery systolic pressure due to pulmonary embolism was reported to be decreased exponentially to near-normal values after an initial dynamic phase within 1 month in more than 90% of patients[14–16]. In the presence of the interatrial shunt flow with right to left direction, it seems reasonable that massive and recurrent thromboembolism originated from deep veins should have been directed to left atrium and systemic circulation through the defect as a result of driving pressure gradient, before arriving at the pulmonary arteries. However, neither history of paradoxical arterial embolism, nor history of acute pulmonary embolism — which may be expected in recurrent and massive venous thromboembolism — was documented in our cases. As a conclusion, thrombus formation within the pulmonary artery due to sluggish flow and/or altered pulmonary vascular physiology is

### Table 1. Patient characteristics.

<table>
<thead>
<tr>
<th>Patients</th>
<th>1</th>
<th>2</th>
<th>3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age/gender</td>
<td>61/male</td>
<td>28/female</td>
<td>49/female</td>
</tr>
<tr>
<td>Haematocrit (%)</td>
<td>58</td>
<td>57</td>
<td>60</td>
</tr>
<tr>
<td>Arterial blood satutation (%)</td>
<td>85</td>
<td>86</td>
<td>84</td>
</tr>
<tr>
<td>Systolic arterial pressure (mmHg)</td>
<td>100</td>
<td>90</td>
<td>95</td>
</tr>
<tr>
<td>Defect</td>
<td>ASD (OS)</td>
<td>ASD (OS)</td>
<td>ASD (OS)</td>
</tr>
<tr>
<td>Diameter</td>
<td>3-6 cm</td>
<td>Patch failure</td>
<td>1.2 cm</td>
</tr>
<tr>
<td>Shunt direction</td>
<td>Bi-directional</td>
<td>Bi-directional</td>
<td>Bi-directional</td>
</tr>
<tr>
<td>RA diameter (cm)*</td>
<td>4.5 ± 5.0</td>
<td>5.0 ± 5.2</td>
<td>5.0 ± 5.4</td>
</tr>
<tr>
<td>RV diameter (cm)*</td>
<td>5.2</td>
<td>4.8</td>
<td>5.1</td>
</tr>
<tr>
<td>RV free wall thickness (mm)</td>
<td>9</td>
<td>8</td>
<td>10</td>
</tr>
<tr>
<td>PASP (mm Hg)</td>
<td>100</td>
<td>70</td>
<td>120</td>
</tr>
<tr>
<td>Diameter of MPA(cm)</td>
<td>3.8-5.2</td>
<td>5.2</td>
<td>5.9</td>
</tr>
<tr>
<td>Diameter of LPA (cm)</td>
<td>4.5</td>
<td>3.9</td>
<td>3.9</td>
</tr>
<tr>
<td>Diameter of RPA (cm)</td>
<td>3.3</td>
<td>2.8</td>
<td>3.5</td>
</tr>
<tr>
<td>SEC</td>
<td>RPA</td>
<td>RPA, LPA</td>
<td>RPA, LUPV, LA</td>
</tr>
<tr>
<td>THR</td>
<td>RPA</td>
<td>RPA, LPA</td>
<td>LPA</td>
</tr>
<tr>
<td>Length (cm)</td>
<td>10</td>
<td>9</td>
<td>8</td>
</tr>
<tr>
<td>Thickness (cm)</td>
<td>1.8</td>
<td>2</td>
<td>2</td>
</tr>
</tbody>
</table>

*Diastolic.

MPA, main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery; SEC, spontaneous echo contrast; THR, thrombus; PASP, pulmonary artery systolic pressure; RA, right atrium; RV, right ventricle; LUPV, left upper pulmonary vein; LA, left atrium; ASD (OS), atrial septal defect (ostium secundum).
considered to be a more plausible explanation than venous thromboembolism.

References


