Childhood Idiopathic Pulmonary Hypertension: a Case Report

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Introduction

Pulmonary arterial hypertension (PAH) is characterized as average pulmonary artery pressure of greater than 25 mm Hg at rest (normal is <14 mm Hg). Childhood idiopathic pulmonary arterial hypertension (CIPAH) is distinguishable from PAH by the absence of family history or underlying disease. Symptoms of CIPAH include dyspnea, exercise limitation, and syncope. Common organ-specific changes include right ventricular hypertrophy and right ventricular strain. Diagnosis for the disease is poor with one, three, and five year survival rates of 68%, 48%, and 35%, respectively.1

In this report, we present the case of a six-year-old girl with idiopathic pulmonary arterial hypertension who died following an exacerbation of her disease, aggravated by inadvertent esophageal intubation.

Case Report

A six-year-old girl was first conclusively diagnosed with pulmonary arterial hypertension approximately 3 years after she exhibited the first signs of any abnormality, which encompassed syncopal episodes on exertion. During the interim, she progressively developed right heart failure. Shortly following the diagnosis, she was readmitted due to seizures secondary to pulmonary hypertensive crises and hypoxia. She was eventually released to home on oxygen therapy.

Two months later, the child’s mother found her daughter seizing and called Emergency Medical Services (EMS). On initial exam, the child was found with a heart rate of 40 bpm and labored respirations, at which time chest compressions were initiated, and respirations were assisted using a bag-valve-mask (BVM) airway. During transport to the emergency department (ED), a second EMS unit met the ambulance on route, and, despite adequate ventilation via the BVM, intubation was attempted. The first attempt was unsuccessful, but a second attempt was reportedly successful. Transport continued to the ED, where resuscitation attempts continued. The patient values.

An additional gross autopsy finding was the identification of an esophageal intubation (Figure 4). The endotracheal tube was found in the patient’s esophagus with the cuff inflated (Fig. 5 & 6) and Blanching underlying mucosa (Figure 7).

Toxicology tests showed therapeutic levels of diazepam. A viral culture was negative.

Figure 1: Autopsy photograph after anterior chest plate removal, showing the enormous heart. Also seen in the right lung. Figure 2: A cross-section of the enlarged heart at autopsy scale in comparison. Note the right ventricular wall (on right side of photograph) is similar in thickness to the left ventricular wall. Also, note the right ventricular cavity is markedly dilated. Figure 5: The markedly congested lungs at autopsy.

The lungs exhibited dark red congestion (Figure 1). The heart was notably enlarged (Figures 4-6) with both the right and left ventricular walls measuring 0.9 cm (Fig. 2).

Crossly, the lungs displayed dark red-brown congested parenchyma (Figure 3).

Table 1: Normal 6-year-old female pulmonary artery values versus patient values.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Normal</th>
<th>Patient</th>
</tr>
</thead>
<tbody>
<tr>
<td>mPAP (mm Hg)</td>
<td>14</td>
<td>72</td>
</tr>
<tr>
<td>PVR (HRU)</td>
<td>&lt;3</td>
<td>21.8</td>
</tr>
<tr>
<td>PCWP (&lt;15 mm Hg)</td>
<td>11 mm Hg</td>
<td>96</td>
</tr>
<tr>
<td>Liver wt. (g)</td>
<td>100-467</td>
<td>120</td>
</tr>
<tr>
<td>Heart wt. (g)</td>
<td>90-197</td>
<td>260</td>
</tr>
<tr>
<td>Height (in)</td>
<td>45</td>
<td>45</td>
</tr>
<tr>
<td>Weight (lbs)</td>
<td>44</td>
<td>45</td>
</tr>
</tbody>
</table>

Discussion

Childhood idiopathic pulmonary arterial hypertension (CIPAH) is characterized by dyspnea (75%), exercise limitation (31%), and syncope (31%) on examination.2 At clinical diagnosis, the patient discussed in this case had a documented history of all three of these symptoms. 80% of patients with CIPAH demonstrate a right ventricular heave; 23% exhibit hepatomegaly, and 8% shows elevated jugular venous pressure. ECG changes indicate right ventricular hypertrophy in 85% of cases and right ventricular strain in 50%.2

Microscopic autopsy findings are indicative of pulmonary hypertension. In the pulmonary arterial hypertension, congestion contributes to a delay onset and chronic dilatation of the vessels lead to narrowing of the vascular lumen. These microscopic changes, as well as medial hypertrophy, are indicative of severe pulmonary hypertension rather than moderate or mild forms of the disease.8

In addition, gross changes to the esophagus were noted, indicating the contributory cause of inadvertent esophageal intubation. Specifically, the cuff of the intubation tube was found inflated in the esophagus, while Blanching of the underlying mucosa was noted at autopsy. Risk factors for esophageal intubation include disease onset and increasing age.9 In a study of critically-ill children, esophageal intubation occurred in 8% and 3% suffered complications.10

Whether this child would have survived this episode if she had not had an esophageal intubation cannot be stated to any degree of certainty. If a forensic pathologist discovers an esophageal intubation at autopsy in a case where death had not yet occurred (via review of the clinical/EMS history), then it is appropriate to consider the esophageal intubation as contributory to death. As per custom, if any non-natural process contributes to a death, the manner of death must be deemed natural, the manner of death ruling is determined by the non-natural process. If the non-natural process contributes to a death, the manner of death ruled as an accident.

The clinical and autopsy findings demonstrate many of the diagnostic pathological findings of childhood idiopathic pulmonary hypertension. Gross findings, for example, marked hepateomegaly and cardiomegaly with dilation of the right ventricle, as well as microscopic findings, such as concentric intimal fibrosis, are indicative of the severe form of the disease that ultimately was considered the major factor in this death. The case demonstrates an example of CIPAH, with especially impressive heart findings. In addition, the case reminds healthcare professionals of the potentially devastating effects of esophageal intubation.

References


