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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 = 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. Prognosis for the disease is poor with one, three, and five year survival rates of 68%, 48%, and 35%, respectively.

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 consecutively 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 she progressively developed right heart failure.

The markedly enlarged heart at autopsy (scale in centimeters). Note that the right ventricular wall (on right side of photo) is thickened (Figure 5). The markedly congested lungs at autopsy (Figure 6).

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 (Figure 5 & 6) and blanched underlying mucosa (Figure 7).

Microscopically, the heart displayed myocyte hypertrophy (Figure 8), and the lungs exhibited intra-alveolar hemosiderin laden macrophages and marked vascular changes (Figure 9), as well as pulmonary arterial intimal cellular proliferation, medial hypertrophy, and concentric intimal fibrosis with intimal narrowing (Figures 10-13). Additionally, pulmonary artery branches were dilated.

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

Discussion

Childhood idiopathic pulmonary arterial hypertension (CIPAH) is characterized by dyspnea (75%), exercise limitation (31%), and syncope (31%) on examination. 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 hypertensive heart, 23% exhibit hepatomegaly, and 8% show elevated jugular venous pressure. ECG changes demonstrate right ventricular hypertrophy in 85% of cases and right ventricular strain in 50%.

Microscopic autopsy findings are indicative of pulmonary hypertension. In the pulmonary circulation, these changes consist of a diminution of distal and intimal fibrosis lead to narrowing of the vascular lumen.8 These microscopic changes, as well as medial hypertrophy, are indicative of severe pulmonary hypertension rather than moderate or mild forms of the disease.10

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 agitation on admission and decreasing age.11 In a study of critically-ill adults, 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 causes contribute to a death, the manner of death is to be declared natural, the manner of death ruling is determined by the non-natural event.12 As such, the manner of death in the presented case was ruled an accident.

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

Autopsy Findings

Gross and microscopic abnormalities indicative of childhood idiopathic pulmonary hypertension were noted at autopsy. The heart was notably enlarged (Figure 1) and exhibited marked right ventricular dilatation and hypertrophy with both the right and left ventricular walls measuring 0.9 cm (Figure 2).

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

Final Diagnosis

Given the autopsy findings and the history as presented, the cause of death was ruled as primary pulmonary arterial hypertension with a contributing factor of esophageal intubation. The following comment was included in the autopsy report: While the underlying severe natural disease may have caused death in and of itself this case, the esophageal intubation cannot be ignored as a contributory cause. The manner of death, therefore, was ruled as accident.

References