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Case Report

Sudden, unexpected infant death due to pulmonary arterial hypertension

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ABSTRACT

A 3-year-old girl with no particular medical history complained of a stomachache and died on the way to the hospital. The autopsy revealed marked right ventricular hypertrophy and dilation with no other cardiac abnormalities. Microscopically, the pulmonary small arteries showed marked medial hypertrophy and varying degrees of intimal and adventitial thickening. We supposed that the cause of death was attributable to pulmonary arterial hypertension (PAH). PAH is a rare disease that can cause sudden, unexpected death at any age. Forensic pathologists should consider PAH in the differential diagnosis of sudden death.

1. Introduction

Pulmonary arterial hypertension (PAH) is a rare disease characterized by high pulmonary vascular resistance and arterial pressure that can lead to right heart failure and death [1]. PAH is a progressive disease with poor prognosis, and sudden death is not unusual [2,3]. PAH occurs mainly in young adult women, but can affect individuals of any age [1,2]. We herein report an autopsy case of PAH in an apparently healthy 3-year-old girl.

2. Case report

A 3-year-old girl complained of a stomachache and shoulder pain at dinner, and these symptoms immediately worsened upon lying down. She was noted to be cyanotic and breathing shallowly when the ambulance arrived. She was taken to the emergency room, but could not be resuscitated. She died approximately 40 min after the onset of the symptoms.

The patient was the firstborn child. One month before her death, she was treated for adenovirus infection. She had a persistent slight cough, but appeared to be well until just before her death.

3. Postmortem radiological findings (Fig. 1)

A chest radiograph revealed enlargement of the cardiac silhouette. Horizontal long-axis computed tomography (CT) showed right ventricular wall thickening and right atrial enlargement. The enlarged right ventricle was also clearly demonstrated on short-axis CT. A lung image showed irregular consolidation around the hilum of the lungs. The bronchovascular bundle was thickened. These findings indicated the presence of pulmonary congestion and edema. A ground-glass opacity was seen in the peripheral region of the lungs. In particular, nodular ground-glass opacities were present, possibly representing inflammation of the bronchi or bronchioles. CT images were reconstructed with soft tissue and lung kernels to provide 1.0-mm slices each.

4. Autopsy findings

The patient's body was 96 cm in height and weighed 13 kg. Apart from some needle marks on her neck, there were no apparent antemortem injuries.

Her heart weighed 112 g, and her right ventricle showed prominent hypertrophy and dilation with a wall thickness of 0.5 cm (Fig. 2). The widths of the aortic valve, pulmonary valve, mitral valve, and tricuspid valve were 3.8, 4.0, 6.5 and 7.5 cm, respectively. The foramen ovale was closed, and we found no other cardiac abnormalities.

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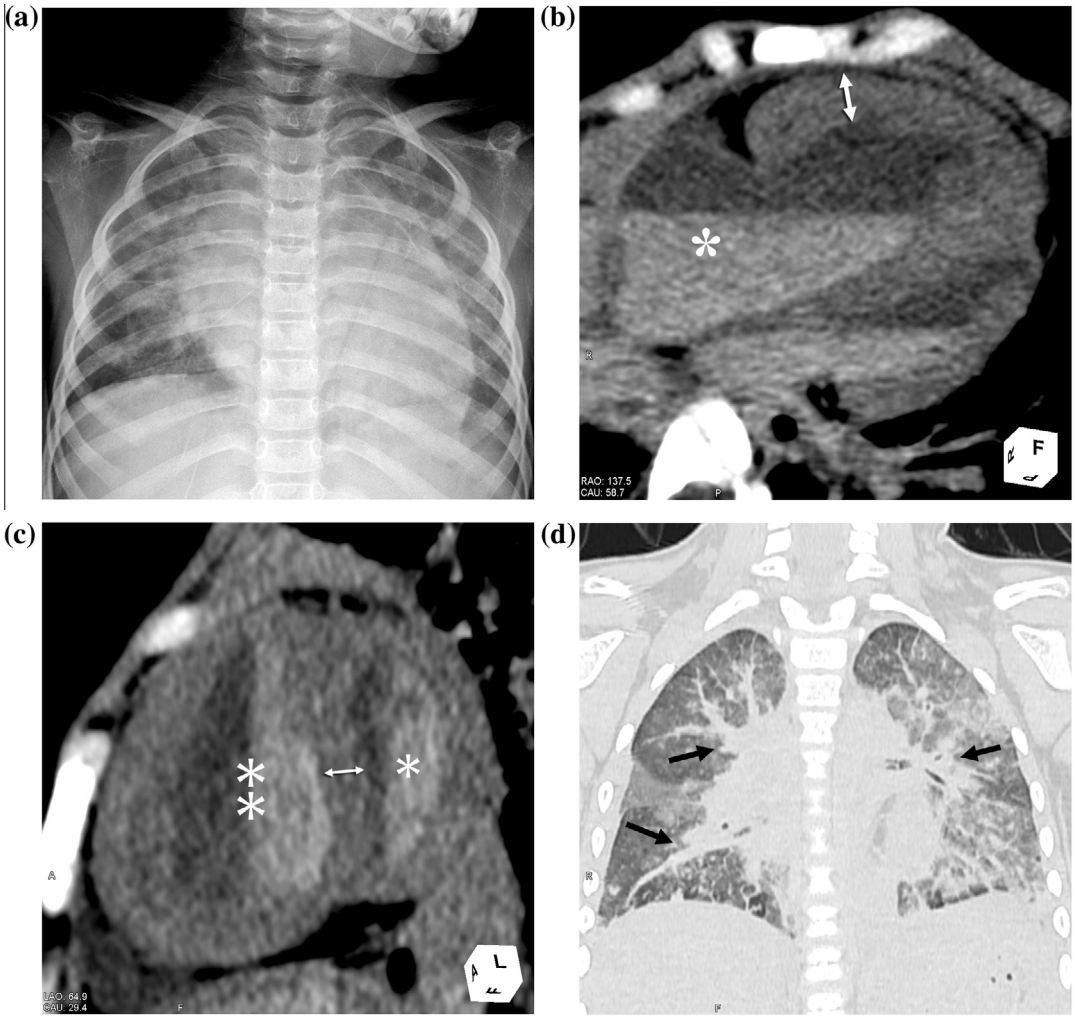

Fig. 1. (a) Supine anteroposterior chest radiograph shows an enlarged cardiac silhouette exceeding 50% of the cardiothoracic ratio. (b) In order to be able to visualize the small density differences within cardiac wall and cavity, quite narrow image window arranged (window width = 90 Hounsfield unit (HU), window level = 50 HU). Horizontal long-axis heart multiplanar reconstruction (MPR) computed tomography (CT) image (i.e., four-chamber view) shows thickening of the right ventricular wall (double-headed arrow), a dilated tricuspid annulus (arrow), and an enlarged right atrium (star). The blood in the heart is separated into two layers due to postmortem hypostasis. (c) Short-axis heart MPR CT image shows an enlarged right ventricle (double asterisk) and thickening of the interventricular septum (double-headed arrow). The left ventricle is normal in size (asterisk). (d) Coronal lung MPR CT image shows consolidation around the bilateral hilar area and a thickened bronchovascular bundle (arrows). A ground-glass opacity is seen in the peripheral region of the lungs.

The patient's lungs (left, 142 g; right, 163 g) were congested on the dorsal side and slightly edematous. No macroscopic stenosis of the pulmonary trunk or either of the main arteries was seen. There were no abnormal findings in the other organs.

5. Histopathology

The lungs showed relatively severe congestion and moderate to severe edema. The pulmonary small arteries showed marked medial hypertrophy and varying degrees of intimal and adventitial thickening. Frequently, many of these arteries were almost completely obstructed by medial hypertrophy and concentric laminar intimal thickening (Fig. 3). Mononuclear inflammatory cell infiltration mainly comprising lymphocytes was seen in and around the bronchial and bronchiolar walls (Fig. 4). These findings were particularly apparent in the right lower lobe. In addition, some peribronchial lymph nodes were swollen and contained germinal centers.

Myocardial cell hypertrophy was seen in the right ventricle. There were no apparent findings indicative of myocardial ischemia. No inflammatory cell infiltration was seen in either ventricle.

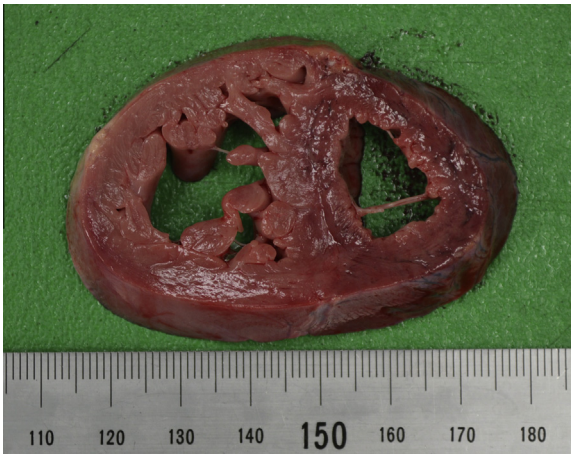

Fig. 2. The right ventricle (left side of figure) is markedly enlarged and its wall is thickened (0.5 cm).