Case #137

NAME Educational Activities Committee

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A 22-month-old female, dichorionic, diamniotic, preterm (33 w) twin, was found dead 15 minutes after being placed down post feeding.
She had a history of plagiocephaly, cleft soft palate, mild fine and gross motor delays, truncal hypotonia, severe obstructive sleep apnea, bilateral middle ear dysfunction, gastroesophageal reflux and a hemodynamically nonsignificant atrial septal defect (ostium secundum), for which she underwent extensive genetic study with no conclusive results.
Postmortem examination showed organs all below normal weight for age, thymus with stress involution, and stage 2 hepatic fibrosis.

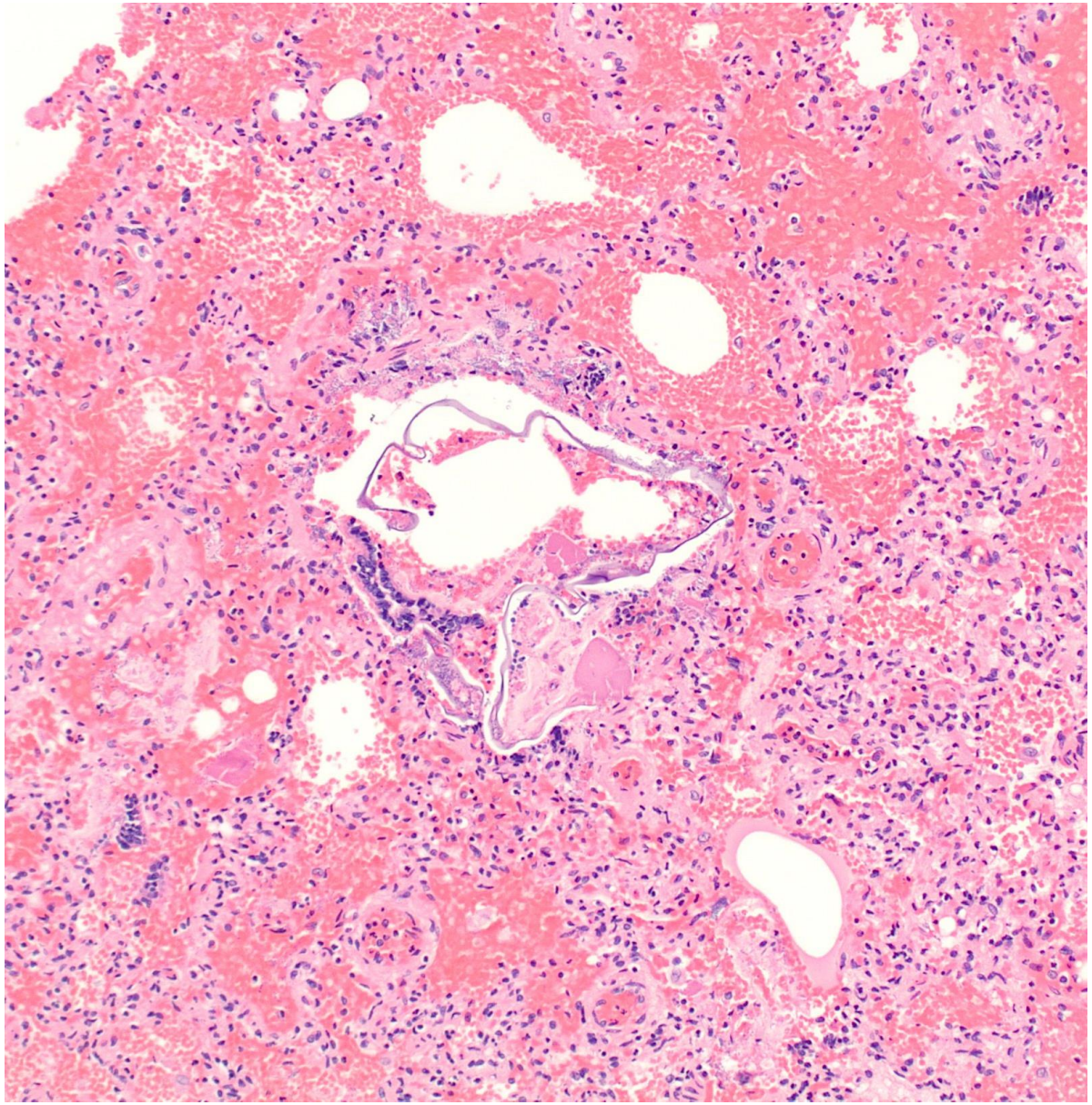
Based on the microscopic images (H&E – 100X), which relevant information has been omitted in the case presentation?

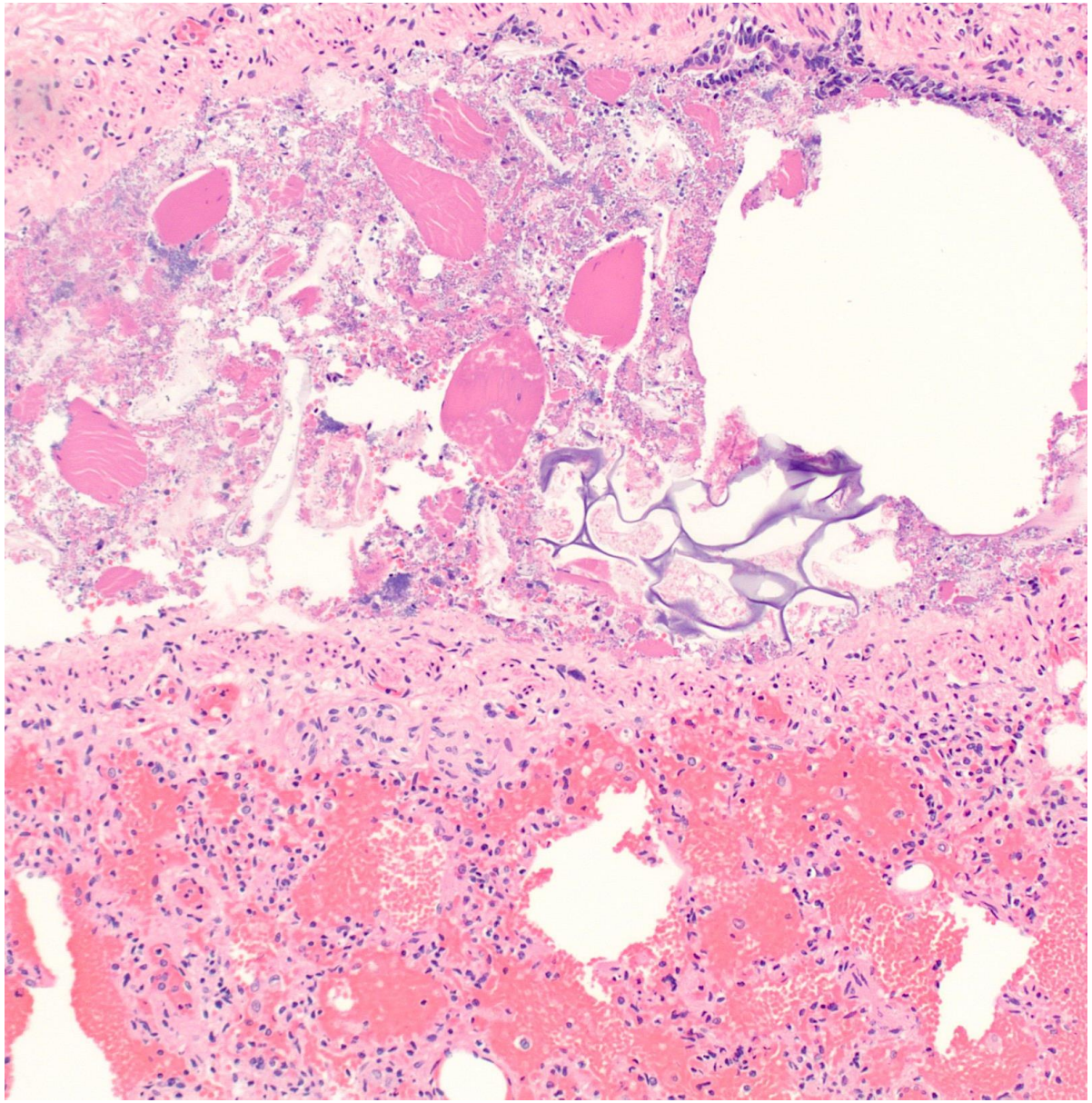
A. She underwent a kidney biopsy showing linear deposits of IgG under direct immunofluorescence.

B. She was brought to ED one month earlier for a rash on the lower extremities.

C. She was followed by a speech therapist and had a gastric tube.

D. A cardiac catheterization procedure was performed with detection of pulmonary arterial hypertension.





CORRECT ANSWER....

C. She was followed by a speech therapist and had a gastric tube (correct – 65.30%)

Aspiration is the inhalation of foreign material into the airways beyond the vocal cords. Pathologic changes in the lungs due to aspiration injury follow a characteristic pattern and include degeneration of the bronchiolar epithelium, pulmonary edema and hemorrhage, focal atelectasis, fibrin exudation, and infiltration by acute inflammatory cells. Food aspiration can cause pulmonary hemorrhage through several mechanisms: direct physical injury, chemical irritation (especially from acidic gastric contents), and severe inflammation, which can sometimes progress to acute respiratory distress syndrome (ARDS). Under normal conditions, gastric contents are sterile. However, changes in gastric fluid pH can create an environment that allows potentially pathogenic microorganisms to thrive. For instance, gram-negative bacteria may colonize the stomach in patients receiving enteral feeding. If gastric aspiration occurs under these conditions, lung infection may result from the bacteria present in the gastric contents.

In the image, vegetable fibers and food particles are visible. Oropharyngeal dysphagia and gastroesophageal reflux can both lead to aspiration. In the presented case, aspiration was a significant risk and contributed to the decision to place a gastrostomy tube (G-tube), through which the infant was being fed.

Other responses...

A. She underwent a kidney biopsy showing linear deposits of IgG under direct immunofluorescence. (incorrect – 7.90%)

This finding is linked to Goodpasture syndrome, an anti-glomerular basement membrane (anti-GBM) disease caused by circulating autoantibodies that target the glomerular basement membrane of capillaries. Goodpasture syndrome is a very rare cause of alveolar hemorrhage in children, who usually also develop rapidly progressive glomerulonephritis. Kidney biopsy remains the gold standard for diagnosis. Light microscopy reveals crescentic glomerulonephritis, while immunofluorescence stains typically show bright linear deposits of immunoglobulin G (IgG) - primarily IgG-1- and complement (C3) on the glomerular basement membrane.

Goodpasture syndrome does not account for aspiration of foreign material in the lungs, and the subject has no history of renal symptoms.

B. She was brought to ED one month earlier for a rash on the lower extremities. (incorrect – 5.15%)

This finding pertains to the clinical presentation of Henoch-Schönlein purpura (HSP), a common leukocytoclastic vasculitis characterized by the deposition of immunoglobulin A (IgA) immune complexes in the small vessels of multiple organ systems. Palpable non-thrombocytopenic purpura with a predominance in the lower limbs is a key criterion for diagnosing HSP, along with at least one of the following: abdominal pain, histopathology showing typical leukocytoclastic vasculitis with IgA deposition (skin biopsy) or proliferative glomerulonephritis with IgA deposition (kidney biopsy), joint symptoms, or renal involvement. Conversely, pulmonary hemorrhage is a very rare complication that carries high morbidity and mortality.

HSP does not explain the presence of foreign material in the lungs, and the subject had no pertinent clinical history.

D. A cardiac catheterization procedure was performed with detection of pulmonary arterial hypertension. (incorrect – 21.65%)

Pulmonary arterial hypertension (PAH), is characterized by an elevated mean pulmonary artery pressure with a pulmonary artery wedge pressure of ≤15 mmHg. Congenital heart defects (CHD) such as ventricular (VSD) and atrial septal defects (ASD) can cause PAH through a left-to-right shunt mechanism that exposes the pulmonary circulation to systemic pressure, resulting in volume overload of the left ventricle, volume/pressure overload in the pulmonary circulation, and possibly life-threatening pulmonary hemorrhage.

Unlike VSD, ASD generally progresses more slowly to severe PAH due to lower atrial pressures. The lack of hemodynamic impact in the presented case makes it unlikely that this is the cause of pulmonary hemorrhage.

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