

goldfinger M

University Hospitals Case Medical Center , Beachwood , Ohio, United states

Introduction:

Leigh syndrome is described as subacute necrotizing encephalomyelopathy. The typical patient (pt.) with this syndrome will have psychomotor retardation, hypotonia, spasticity, chorea, ataxia, and peripheral neuropathy. Pulmonary hemorrhage (PH) is not a feature of Leigh syndrome.

Case presentation:

The pt. is a 6 year old, 11 kg male who presents to the operating room (O.R.) for direct laryngoscopy/bronchoscopy (DLB) and nasal endoscopy secondary to epistaxis and hemoptysis. His past medical history is significant for prematurity (27 weeks), Leigh syndrome, mitochondrial disease, GERD with chronic lung disease secondary to aspiration pneumonia, right sided pneumatocele, developmental delay, failure to thrive, blindness, hypothyroidism, hepatosplenomegaly, CD4 deficiency and ectodermal dysplasia. The pt. was transferred from the Pediatric Intensive Care Unit (PICU) to the O.R.. Routine monitors were in place. The pt. was induced with Etomidate, Fentanyl and <1% Sevoflurane while breathing 100% oxygen via face mask. The head of the bed was then turned toward the otolaryngologist. The surgeon performed a DL and intubated the trachea with a 4.5 mm microcuffed endotracheal tube (ETT). Copious amounts of bright red blood was immediately noted from the ETT. It became difficult to ventilate the pt..The ETT was suctioned and epinephrine (Epi) delivered down the ETT in an attempt to obtain hemostasis. A pulmonologist performed a flexible bronchoscopy through the ETT and did not identify the source of pulmonary hemorrhage. Bradycardia secondary to hypoxia ensued and the patient was ventilated with 100% oxygen, chest compressions performed and intravenous (IV) Epi, atropine, vasopressin, calcium chloride and sodium bicarbonate administered. Copious blood repeatedly came through the ETT. PRBCs, FFP, platelets and factor 7 were administered IV. A subsequent bronchoscopy revealed a clot above the carina. The patient was brought back to the PICU intubated on an Epi. and Vasopressin infusion.

That evening the pt. underwent a cardiac cath. to identify and perhaps coil the blood vessel responsible for the PH. A source was not identified. The pt. suffered a subsequent cardiac arrest in the PICU and has spent a protracted course of 8 weeks thus far in the PICU. Although he is extubated on CPAP and has not had a recurrence of PH, he did suffer from ischemic hypoxic encephalopathy.

Discussion:

This case demonstrates a case of a boy with numerous medical issues who presented with and was treated for PH. Interestingly, there have been numerous cases of PH in infants and children living in the Cleveland, Ohio, USA, area described by various investigators[1]. Some of these pts. were exposed to a fungus, *Stachybotrys chartarum* endemic to homes with water damage in the Cleveland area. The incidence has declined significantly in recent years. Our patient, a foster child living in the Cleveland area, had bronchoalveolar lavage samples which were negative for fungus. Although the etiology of this pts. PH is unclear, one possibility is an infected pneumatocele.

References:

1. Tripi PA, Modlin,S.,Sorenson,WG,Dearborn,DG, Acute pulmonary hemorrhage in an infant during induction of general anesthesia. Paediatric Anaesthesia 2000;10: 92-94.
