Autopsy Case Reports

Image in Focus

Kernicterus

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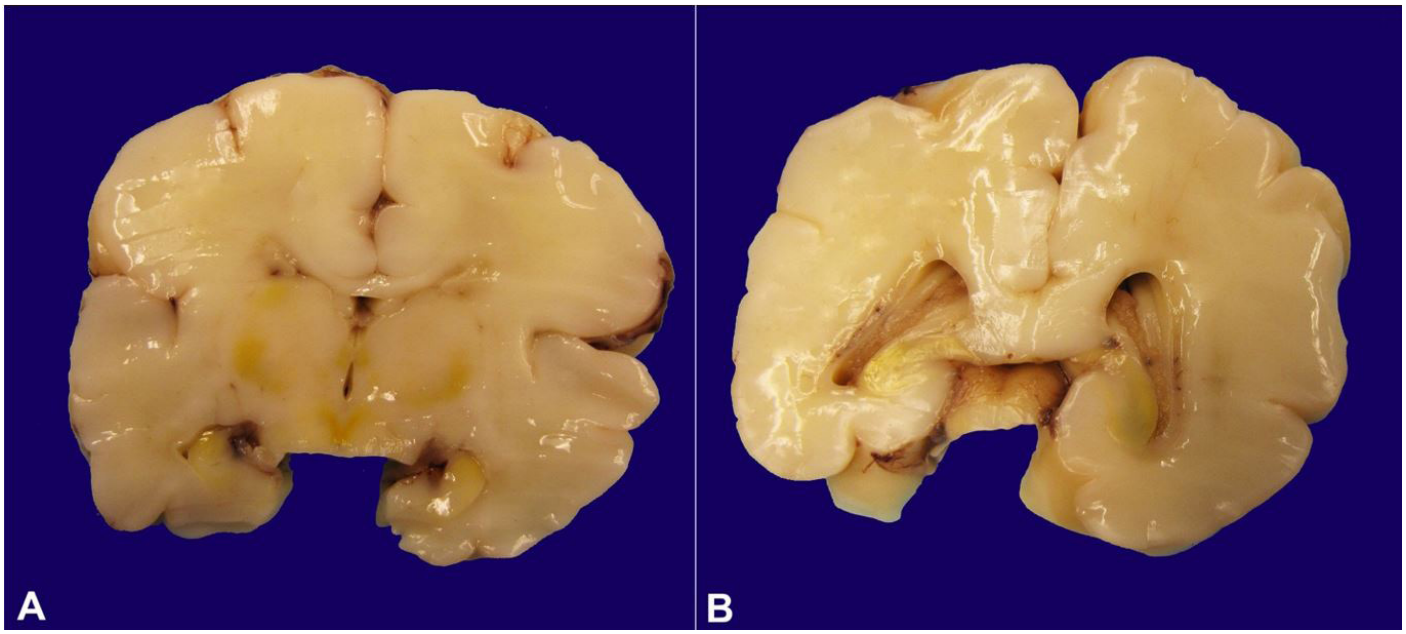

Figure 1. Macroscopic cross section of the brain demonstrating extensive yellow discoloration in the basal ganglia, thalamus, cerebellum, tegmentum **(A)** and floor of the ventricles **(B)**. Lateral ventricles lined by germinal matrix and demonstrating minute hemorrhages **(B)**.
Image courtesy Dr. Ameer Hamza

Kernicterus is a bilirubin-induced brain damage most commonly seen in infants. Regions of the brain most commonly affected are the basal ganglia, hippocampus, geniculate bodies and cranial nerve nuclei.[1] The exact incidence of kernicterus is unknown; however, most recent data from the United Kingdom and Canada suggests kernicterus occurring at a rate of 1 to 2 in 100,000 live births.[2,3] The risk of developing kernicterus increases considerably in infants with bilirubin levels >25 mg/dL while levels >30 mg/dL are associated with extremely high risk and irreversible damage.[4] Any event that leads to increased bilirubin production or decreased elimination can lead to hyperbilirubinemia and thus kernicterus. These include, but are not limited to, polycythemia, hemolysis due to Rh isoimmunization and congenital inherited defects of enzymes involved in bilirubin metabolism. Additionally, systemic factors such as hypothyroidism, certain drugs and infections particularly meningitis increase the risk of kernicterus. The clinical presentation can

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be divided into three phases. In phase 1, the infant demonstrates decreased alertness, hypotonia, and poor feeding. This is followed by a phase of hypertonia of the extensor muscles. Progress to this phase invariably leads to long-term neurologic deficits. Phase 3 usually occurs in infants aged >1 week, and they typically demonstrate hypotonia. Work-up includes blood indices, comprehensive metabolic panel and quantitative measurement of total and direct bilirubin. Neuroimaging is of limited value, however, it can help rule out other diagnoses. The definitive treatment is removing bilirubin from the blood via exchange transfusion.

From surgical pathology standpoint, a brain biopsy is rarely done to make a diagnosis. Historically, the term "kernicterus" referred to an anatomic diagnosis made at autopsy based on a characteristic staining of basal ganglia, observed in infants who died with marked hyperbilirubinemia. The neuronal changes include pyknotic nuclei, cytoplasmic vacuolation, and loss of the Nissl substance.

The Figure refers to the gross appearance of the brain in a deceased 16-day-old infant born at 29 weeks of gestational age via normal spontaneous vaginal delivery to a G4 P3 mother with a history of pre-eclampsia and eclampsia in previous gestations, who did not receive prenatal care and tested positive for tetrahydrocannibinol on urine drug screen. After birth, the infant was noted to be in respiratory failure, was placed on positive pressure ventilation and transferred to the neonatal intensive care unit (NICU). NICU stay was notable for multiple episodes of apnea, bradycardia, desaturations and persistent hyperbilirubinemia with bilirubin levels up to 31 mg/dl. Despite intensive phototherapy, the patient became more jaundiced and had feeding difficulties. Double volume exchange transfusion was planned and the patient had an IV catheter inserted by pediatric surgery; however, he went into cardiopulmonary failure and was pronounced dead.

The detailed brain examination at autopsy revealed extensive yellow discoloration in the basal ganglia, thalamus, cerebellum, tegmentum, and floor of the ventricles (Figure 1A). The lateral ventricles were lined by germinal matrix and demonstrated mild hemorrhage (Figure 1B).

Keywords
Kernicterus; Bilirubin; Infant, Newborn, Autopsy.

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The case is from St. John Hospital and Medical Center, Detroit, MI, USA and is in accordance with the recommendations of institutional review board.

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