H118 Multifocal Intracranial Hemorrhage in Congenital Neurosyphilis: Autopsy Findings and Literature Review

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Learning Overview: The goal of this presentation is to review the common clinicopathologic and autopsy findings of congenital neurosyphilis and discuss an uncommon presentation of the disease.

Impact on the Forensic Science Community: This presentation will impact the forensic science community by increasing awareness of congenital neurosyphilis and its manifestations.

Background: Congenital syphilis occurs by transplacental transmission of the spirochete *Treponema pallidum* from an infected pregnant woman to her fetus. Infection results in a spectrum of clinical manifestations, ranging from asymptomatic infection to sudden death.[1] The central nervous system is rarely affected in infants of treated mothers; however, if untreated, ongoing dissemination of the organism can lead to neurosyphilis.

Autopsy: A male infant was born at 29 weeks gestational age via cesarean section for fetal bradycardia. The pregnancy was complicated by intrauterine growth restriction and maternal history of syphilis and alcohol abuse. There were minimal signs of life at delivery, and despite resuscitative efforts, the infant passed away within an hour of birth.

Autopsy revealed a developing and normally formed infant male. The face and chest were involved by a tan-purple maculopapular rash. Small pleural and peritoneal effusions and minimal thymic hemorrhage were present, consistent with resuscitative efforts. An isolated ostium secundum atrial septal defect was identified. Bacterial and viral cultures of the lungs and cerebrospinal fluid were negative, and cultures of the blood showed mixed growth consistent with postmortem overgrowth.

Neuropathologic examination demonstrated a large subdural hematoma overlying the cerebral convexities. Scattered subarachnoid hemorrhage was present, involving the isocortex, brainstem, and spinal cord. Lymphoplasmacytic and histiocytic predominant inflammation involved the meninges. The subcortical white matter and spinal cord demonstrated severe white matter necrosis, edema, and petechial hemorrhage. The lateral ventricles were enlarged. Immunohistochemical staining for spirochetes within the parenchyma of the frontal lobe and cingulate gyrus was positive.

The cause and manner of death were certified as congenital neurosyphilis, natural.

Discussion: Congenital neurosyphilis generally manifests as syphilitic meningitis, with early (newborn) and late (1–2 years old) manifestations of the disease.[2] Despite being an acute bacterial infection, syphilitic meningitis is characterized by a mononuclear inflammatory infiltration of the leptomeninges, comprised of plasma cells, lymphocytes, and macrophages. Extension of the cellular infiltrates into brain parenchyma through Virchow-Robin (perivascular spaces), or direct extension into superficial cortex, has been suggested for those with intraparenchymal involvement.[3]

Syphilitic meningitis often progresses to chronic meningovascular syphilis (syphilitic vasculitis) in untreated cases. Inflammation and reparative changes cause cranial nerve palsies and hydrocephalus; over time, the vasculitis may lead to thrombosis, ischemia, and infarction.[4,5] Aneurysm formation and rupture with cerebral hemorrhage can occur; however, this has not been described in premature infants.

Multifocal intracranial hemorrhage represents a novel manifestation of congenital neurosyphilis, an already uncommon disease. The intracranial hemorrhage in this case has numerous potential syphilitic-mediated etiologies.[6] Intracranial hemorrhage has been described in infants with non-syphilitic acute bacterial meningitis and central nervous system vasculitides. Moreover, spontaneous intracranial hemorrhage is a known complication of Disseminated Intravascular Coagulation (DIC), which has rarely been associated with congenital syphilis.[7] Syphilis should be considered in the differential for disease processes that cause meningitis with associated hemorrhage.

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