H77 Forensic Neuropathology of Cerebral Palsy (CP): The Implications for Cause-of-Death Determination

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After attending this presentation, attendees will better understand the spectrum of neuropathologic substrates underlying CP.

This presentation will impact the forensic science community by informing attendees of: (1) the differential diagnosis of CP, including perinatal hypoxia-ischemia, developmental anomalies, post-infectious sequelae, and storage/metabolic/genetic disorders; (2) the gross and microscopic presentations of these conditions; and, (3) the approaches for forensic neuropathologic workup at autopsy.

Introduction: CP is the most prevalent physical disability in childhood and includes a heterogeneous constellation of non-progressive movement and posture disorders due to insults to the developing fetal or infant brain. Symptoms are spasticity (hemiplegia, diplegia, quadriplegia), dyskinesia, ataxia, and/or hypotonia, as well as variable cognitive impairment. Colloquially, "cerebral palsy" is applied to many ill-defined neurologic conditions arising in childhood and leading to long-term institutionalization. Thus, medical examiners often investigate their deaths and are responsible for determining the underlying cause and manner of death. This study sought to more accurately categorize the underlying neurolopathologic substrate in cases of "CP" referred to the City of New York Office of Chief Medical Examiner (OCME) over a two-year period.

Methods and Results: During this interval, 20 cases were referred to the OCME with the clinical diagnosis of CP. The investigation report, medical documentation, and clinical history were reviewed, as were gross and microscopic data. Fifteen decedents were male and five female; age range was 2-59 years (median, 28 years). Neuropathology fell into five specific categories: (1) hypoxic-ischemic injury (n=9); (2) genetic/syndromic/metabolic disorders (n= 5); (3) post-infectious sequelae (n=3); (4) traumatic brain injury (n=1); and, (5) other (n=2). For Category 1, examples include perinatal hypoxic-ischemic injuries with periventricular leukomalacia, germinal matrix hemorrhage and periventricular hemorrhagic infarct, status marmoratus, and aqueduct stenosis with hydrocephalus. Category 2 included Coffin-Lowry, Cornelia de Lange, and Klippel-Feil syndromes, and neocerebellar aplasia/hypoplasia.

Neuropathology was directly relevant to cause and manner of death in 15 cases owing to severe disability with general deconditioning, septic and respiratory complications, and malfunctioning of life-support equipment. In occasional cases, the neuropathologic diagnosis had direct impact on an affected family member for whom a specific diagnosis had not been made (e.g., neuronal ceroid lipofuscinosis in a twin).

Conclusion: In virtually all cases labeled as "CP," a specific neuropathologic diagnosis is possible, allowing more accurate categorization of cases and of corresponding cause and manner of death. Such specificity may have important clinical consequences for families and caretakers.

Cerebral Palsy, Hypoxic-Ischemic Injury, Developmental Neuropathology

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