Subacute Sclerosing Panencephalitis: the Devastating Measles Complication is More Common than We Think

Background: Subacute sclerosing panencephalitis (SSPE) is a fatal complication of measles. Thought to be rare, SSPE incidence decreased with routine measles vaccination, but infants with measles remain at highest risk of this complication. We reviewed SSPE cases in California from 1998-2016 to understand current risk factors for SPPE.

Methods: SSPE cases had a clinically compatible illness and either 1) measles IgG antibody detection in the cerebrospinal fluid; 2) characteristic pattern on electroencephalography; 3) typical histologic findings in brain biopsy; or 4) medical record documentation of SSPE-related complications. Cases were identified though a state death certificate search, reports from the Centers for Disease Control and Prevention, or through investigations for undiagnosed neurologic disease. Measles IgG detection was performed using indirect enzyme immunoassay at the California Department of Public Health (CDPH) or by immunofluorescence assay at clinical laboratories.

Results: Seventeen SSPE cases were identified. Males outnumbered females 2.4:1. Twelve (71%) cases had a clinical history of a febrile rash illness compatible with measles; all 12 had illness prior to 15 months of age and measles vaccination. Eight (67%) children were living in the United States when they had measles. SSPE was diagnosed at a median age of 12 years (range 3-35 years), with a latency period of 9.5 years (range 2.5-34 years). Many cases had long-standing cognitive or motor problems prior to diagnosis. Among measles cases reported to CDPH during 1988-1991, incidence of SSPE was 1:1367 for children < 5 years, and 1:609 for children < 12 months at time of measles disease.

Conclusion: SSPE cases in California occurred at much higher rate than previously published among unvaccinated children who were infected with measles in infancy. Protection of infants younger than 12-15 months of age, when measles vaccine is routinely administered, requires avoidance of travel to endemic areas, or early vaccination prior to travel. Clinicians should be aware of the possibility of SSPE in patients with compatible symptoms, even in older patients with no specific history of measles infection. SSPE demonstrates the high human cost of “natural” measles immunity.

Kristen Wendorf, MD, MS[1], Kathleen Winter, MPH, PhD[1], Kathleen Harriman, PhD, MPH, RN[1], Jennifer Zipprich, MS, PhD[1], Robert Schechter, MD[1], Jill Hacker, PhD, MPH[2], Chris Preas, BA[2], James D. Cherry, MD, MSc, FIDSA[3] and Carol Glaser, DVM, MPVM, MD[4], (1)Immunization Branch, California Department of Public Health, Richmond, CA, (2)Viral and Rickettsial Disease Laboratory, California Department of Public Health, Richmond, CA, (3)Pediatric Infectious Diseases, University of California, Los Angeles, David Geffen School of Medicine, Los Angeles, CA, (4)Kaiser Permantente, Oakland, CA

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