P-022 - REYE-LIKE SYNDROME TRIGGERED BY SYNCYTIAL VIRUS INFECTION IN TWO PATIENTS WITH FATTY ACID OXIDATION DISORDERS


Hospital Pablo Tobón Uribe. Medellín – Colombia. leomunera@yahoo.com Link Diagnostico Digital. Medellín – Colombia. Dinamica. Medellín - Colombia

Reye syndrome is a rare but dangerous emergency that children suffer. The etiology and pathogenesis of the Reye syndrome is not clear. Today, special attention is paid to the development of so-called Reye-like syndromes in the context of metabolic defects. Defects of the fatty acid oxidation (FAO) often present in infancy as the metabolic demands of the developing brain are higher in the young brain, mainly due to its vulnerability to hypoxia, hypoglycemia and hypoketonemia. These disorders are often clinically and biochemical silent, but become manifest under specific stressors such as prolonged fasting, fever with infection, mild to moderate exercise and cold exposure. Recurrent hypoglycemia, hypoketonemia, hepatic encephalopathy and microvesicular steatosis of liver are by definition the classical signs of Reye-like syndrome and the pathological signature of FAO disorders. Here, we report on two patients with Reye-like syndrome due to FAO disorders triggered by Syncytial Virus Respiratory infection (SVR). Patient 1, is the first child of non-consanguineous parents, who at 8 months of age began with hypoglycemia, hyperammonemia, hepatomegaly with hepatocellular dysfunction, steatohepatitis and hypertrophic cardiomyopathy secondary to a SVR infection. Basic biochemical tests as well as plasma acylcarnitines analysis by gas chromatography-mass spectrometry (GC-MS) were performed. Patient 2, is the first child of non-consanguineous parents, who in the first month of life began with hypoglycemia. Afterwards hyperammonemia, hepatomegaly with hepatocellular dysfunction, steatohepatitis and hypertrophic cardiomyopathy and myopathy were detected secondary to a SVR infection. Basic biochemical tests as well as acylcarnitines analysis by GC-MS were performed. Both initial biochemical tests showed increase in AST and ALT with hyperammonemia and hypoketotic hypoglycemia. Liver ultrasound demonstrated stenteatohepatitis and the echocardiography, hypertrophic cardiomyopathy. Plasma acylcarnitine analysis demonstrated a significant elevation of C6-C10 acylcarnitines in patient 1 typical of Medium-Chain Acyl-Coenzyme A Dehydrogenase deficiency; in patient 2, a rise in C14: 1, C14: 2, C14, and C12: 1 was seen, suggestive of Very Long-Chain Acyl-Coenzyme A Dehydrogenase deficiency. The description of these two cases confirmed the presence of Reye-like syndrome secondary to SRV infection in patients with FAO disorders in Latin America, highlightening SRV as a new trigger of this disorder.