Submitted by: Caroline Galeotti

Title: Idiopathic multicentric Castleman disease associated with Duchenne dystrophy and cutaneous leukocytoclastic vasculitis in a 4-year-old child

Author: Charlotte Borocco, Claire Meyer, Glory Dingulu, Stéphanie Franchi-Abella, Charlotte Mussini, Isabelle Koné-Paut, Oanez Ackermann, Caroline Galeotti

Summary: Idiopathic multicentric Castleman disease (iMCD) is a rare lymphoproliferative disease characterized by multifocal lymph nodes, severe inflammatory manifestations and overproduction of interleukin 6 (IL-6). A 4-year-old boy, with non-explored behavioral disorder, consulted for an abdominal distension, a poor general state and a low-grade fever for one month. Clinically, he suffered from acute malnutrition, had a waddling walk, an incomprehensible language and a 6 cm hepatomegaly. There was an inflammatory syndrome, a hepatic cytolysis, increased CPK, a hypergammaglobulinemia and anti-smooth muscle antibodies. CT-Scan showed hepatomegaly, a dilatation of the biliary tract and lymph nodes at the hepatic hilum. Lymph node biopsy showed specific characteristics of Castleman disease. Duchenne muscular dystrophy was diagnosed thanks to muscular biopsy and genetic analysis. The patient responded well to tocilizumab, an anti-IL-6 receptor monoclonal antibody. When tocilizumab infusions were spaced out, he developed episodes of cutaneous leukocytoclastic vasculitis with inflammatory digestive involvement. Tocilizumab infusions were brought closer together, every two weeks, and he is receiving enteral nutrition. This observation is the first to describe a boy with an iMCD associated with a Duchenne muscular dystrophy, an inflammatory disease of the digestive tract, and a cutaneous leukocytoclastic vasculitis.