Fulminant Epstein Barr Virus Encephalitis—Can it be Due to X-linked Lymphoproliferative Disease

I read with great interest the recently published article "Fulminant Epstein Barr Virus Encephalitis" [1]. Infectious mononucleosis (IM) is usually a common benign illness caused by Epstein Barr Virus (EBV). I fully agree that neurological manifestations in IM are extremely rare. It may be the presenting feature or occur as a result of complication usually appearing after 1-3 weeks of illness. The authors described two such cases with neurological manifestations as presenting feature in absence of classical signs of IM like sore throat, skin rash, lymphadenopathy, hepatomegaly, splenomegaly. I feel the authors should have considered the background illness *i.e.* X-linked lymphoproliferative (XLP) disease referred to as Duncan disease, a combined immunodeficiency disease that manifest after exposure to EBV. Boys with XLP mutation remain clinically well before exposure to EBV. Exposure to EBV results in fulminant and fatal IM in 60% of cases [2]. The fulminant course ultimately leading to death within 8 weeks after onset of IM in 95% of cases [3]. The fulminant nature of the course of EBV infection in both the cases should draw attention to the possibility of XLP disease. This may also be further supported by the fact that both the cases were boys. There is marked impairment in production of antibodies to the EBV nuclear antigen (EBNA),where as titres of antibodies to viral capsid antigen (VCA) have ranged from absent to markedly elevated [4]. Authors have mentioned that in first case EBV serology was positive in blood and in second case IgM EBV was positive in blood. It is not clearly understood what type of serology was undertaken. Although patients with XLP disease have normal number of T lymphocyte CD 8^+ T cell predominate with inversion of normal CD 4 – CD 8 ratio [3].CD 4^+, CD 8^+ lymphocyte count and its ratio study also could have been done in the index cases. Genetic study for detection of SAP (for SLAM *i.e.* signaling lymphocyte activation molecule.- associated protein) now known as SH2D1A may be done if facilities permit [4].

JB GHOSH
Consultant Pediatrician,
Ex-Professor and HOD, IPGME and R, Kolkata,
Ushashi Housing Society, 245 Vivekananda Road,
Kolkata 700 006, West Bengal.
jbghosh@yahoo.com

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