Probable case of CAEBV associated in evolution with a MS-like syndrome in a 12 year old girl

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Rare patients infected with EBV (Epstein-Barr virus) develop a life-threatening condition termed Chronic active Epstein-Barr infection.

Chronic active Epstein-Barr infection (CAEBV) =

progressive lymphoproliferative disorder with a duration of > 6 months, characterized by:

- markedly elevated levels of antibody to EBV /or EBV DNA in the blood
- infiltration of tissues with lymphocytes
- The absence of any other immunosuppressive condition
- elevated EBV-DNA ,RNA or protein in affected tissues

The only treatment reported successful in literature is **allogeneic HSCT** (hematopoietic stem cell transplantation) .

We present a rare case of a **12 year old** girl, with a history of:

Age

- **6 month** – Acute EBV mononucleosis with **hemofagocitic lymphohistiocytosis** and **fulminant acute liver failure**.

- **3 years** - Abdominal CT show **enlarged liver and spleen**, no adenopathies ;
+ Liver biopsy $\Rightarrow$ Chronic viral hepatitis.

- **4 years** – She starts presenting motor deficit cerebral CT- right and temporal myelination deficit

cerebral MRI - disseminated infratentorial and supratentorial inflammatory lesions.

- **5 years** - Spastic tripareisis , strabismus bilateral VI nerve palsy, swallowing difficulties, gait disturbance, dysarthria – treated with steroids; mild sequelae;

- **10 years** - cerebral MRI for neurological monitoring (2015): sequelae of demyelination lesion; the largest one -left temporal (clinically – no new sign).

- **11 years (August)** : right hemiparesis affecting mostly the right arm -head and cervical spinal cord MRI: new demyelinating lesions both encephalic and spinal cord - favorable outcome with i.v steroid pulse therapy and then 9 months orally;

- **11 years** (January) - cerebral MRI and cervical spinal cord – reduced preexisting demyelinating lesions; new gadolinophilic and hemosiderin lesions in the cervical spinal cord

- **12 years-** walking difficulties – cerebral and spinal cord MRI - important extension of the lesions

Clinical and paraclinical examination show:

Constant:

- Markedly elevated levels of antibodies to EBV in the blood
- Hepatosplenomegaly
- WBC around 3000/mm³
- Low platelet number
- Mild anemia
- Moderate increase of liver enzymes
- Negative oligoclonal bands in CSF.

In conclusion, in the evolution of an infection with EBV, CAEBV may develop with the possibility of further **neurological MS-like syndrome** being associated.