



ANTI-HU ASSOCIATED ENCEPHALITIS: AN INITIAL PRESENTATION OF NEUROBLASTOMA

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17th INTERNATIONAL CHILD
NEUROLOGY CONGRESS

INTRODUCTION

Anti-Hu associated paraneoplastic syndromes are extremely rare and commonly related to a past history of neuroblastoma in childhood.

Anti-Hu-associated paraneoplastic encephalitis (PNE) may be the first manifestation of neuroblastoma.

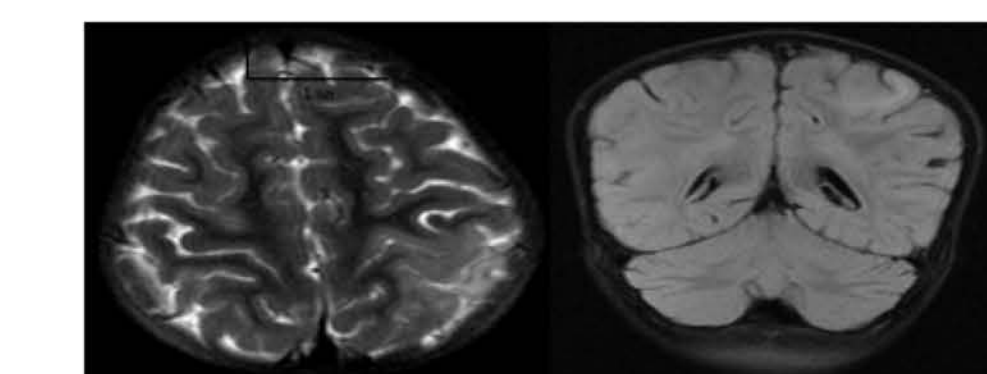
CASE PRESENTATION

A two-year-old girl presented with a focal motor seizure and segmental myoclonus involving the right arm and trunk. Neurological examination revealed Irritability and mild ataxia.

CLINICAL EVALUATION

- **Electroencephalogram (EEG)** at admission showed no ictal activity, but interictal sharp wave discharges in the left central region.
- **Brain Magnetic Resonance Imaging (MRI)** revealed a T2-hyperintense lesion compatible with encephalitis on the left parietal lobe.
- **Spinal MRI** showed a left suprarenal mass, suggesting neuroblastoma.
- **Neuron-specific enolase** was 32 µg/L (normal<15).
- **Cerebrospinal fluid (CSF) Immunoglobulin G index** was high.
- **Oligoclonal bands** in serum and CSF were present.
- The case was initially diagnosed with the **opsoclonus-myoclonus-ataxia syndrome (OMAS)** secondary to neuroblastoma.
- However, the detected parietal lesion raised the suspicion of **paraneoplastic encephalitis**.
- Serum and CSF analysis revealed high levels of **anti-Hu antibodies**.

TREATMENT AND CLINICAL COURSE



1a. Hyperintense signal in left parietal cortex on T2W FLAIR

Encephalitis: Antiviral, antibacterial therapy and IVIg

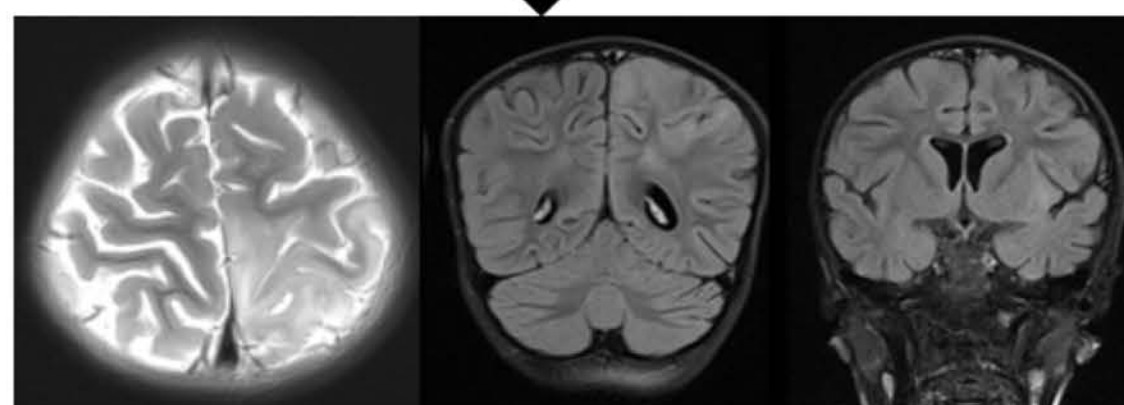


Paraneoplastic encephalitis?

CSF Anti HU +++

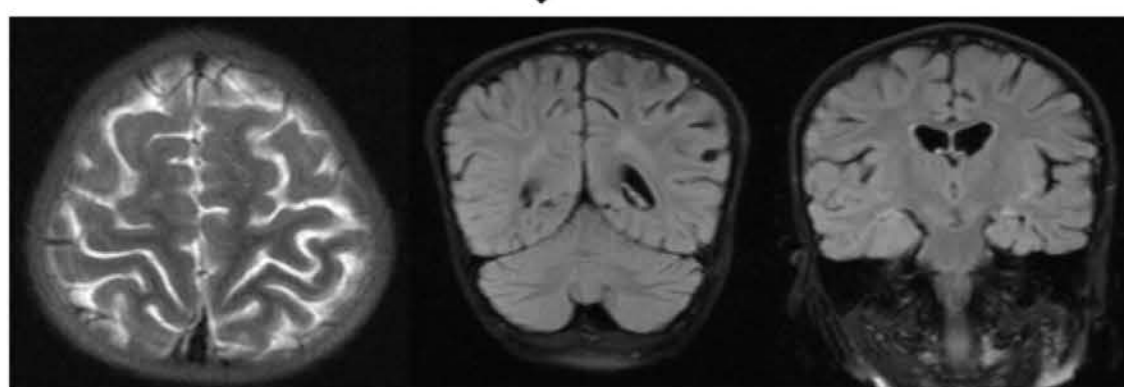
1b. Mass on the anterior surface of the left kidney (neuroblastoma?)

Paraneoplastic encephalitis: Surgery + Dexamethasone

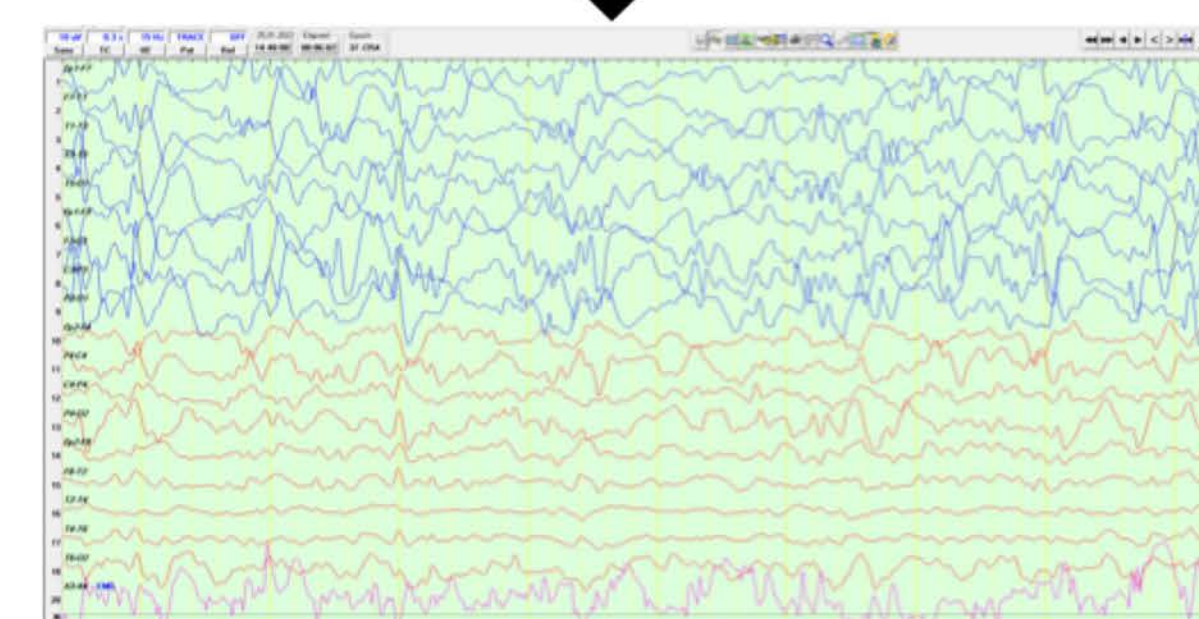


1c. Progression in hyperintensity on left parietal cortex and insular cortex involvement

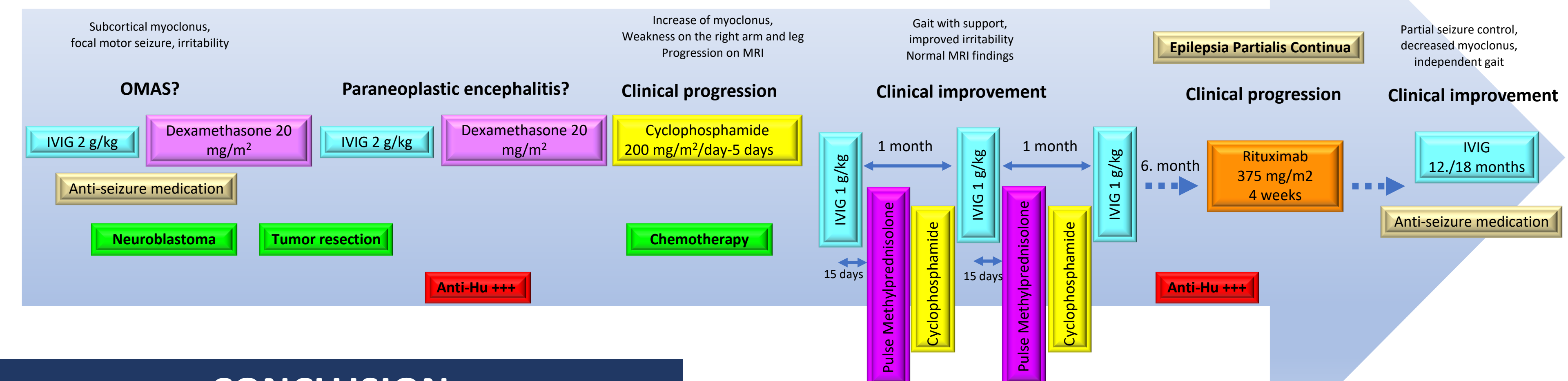
Pulse methylprednisolone + IVIG + Cyclophosphamide / Monthly



1d. Normal brain MRI findings



1e. Spike/polyspike and wave discharges on left hemisphere



CONCLUSION

- Anti-Hu associated encephalitis may also be the first manifestation of neuroblastoma, in addition to OMAS.
- Anti-Hu associated encephalitis has been reported in a small number of pediatric cases, with all having a past history of OMAS and neuroblastoma.
- Epilepsia partialis continua (EPC) has rarely been documented in pediatric patients with anti-Hu mediated encephalitis.
- Aggressive immunotherapy protocols may provide a successful control of EPC.
- In patients with atypical neuroblastoma with/or OMAS, anti-Hu associated encephalitis should be kept in mind.

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