MAGNETIC RESONANCE IMAGING PATTERNS IN PEDIATRIC CENTRAL NERVOUS SYSTEM HEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

Yasser Mazloum Zakareya, Ahmed Adel ElBeheiry, *Marwa Saeid AbdelMaksoud, Basant Gamal Ali Negm

Department of Radiodiagnosis and Intervention, *Department of Pediatric, Faculty of Medicine, Alexandria University

Introduction

HLH is a rare, life-threatening pediatric condition characterized by uncontrolled hyperinflammation caused by various hereditary or acquired immune deficiencies. It is classified into two subgroups: Primary HLH including familial HLH (FHLH) and immunological deficits and secondary HLH which can be caused by infection, cancer, rheumatology, or iatrogenic factors (transplantation, immunological suppression, immune stimulation). HLH is always fatal unless treatment results in a partial or total clinical response. Chemo-immunotherapy with steroids, etoposide, and cyclosporine, as well as rabbit antithymocyte globulin, cyclosporine, and prednisone, can be used with or without intrathecal methotrexate. So far, the only curative therapy strategy is allogeneic hematopoietic stem cell transplantation. Central nervous system (CNS) involvement, has been documented as an independent risk factor for mortality in children with HLH. Clinically, due to the rarity of pathological studies of HLH, making the diagnosis of CNS involvement difficult, clinicians must rely on clinical manifestations combined with imaging and laboratory tests to confirm such diagnosis.

Aim of the work

The aim of this study was to define the distinct MR brain imaging patterns in children with CNS-HLH involvement.

Patient and Methods

This retrospective study was conducted on children diagnosed with CNS-HLH managed at Alexandria University Children's Hospital (AUCH). Data of children admitted within the period from October 2022 to December 2023 were recruited and brain imaging studies were collected and re-evaluated. The brain imaging studies were acquired on a variety of MRI scanners. MR images were included if the following sequences were routinely done: Axial or 3D T1-weighted sequences, Fast spin echo (FSE) T2- weighted axial, coronal and sagittal sequences, Axial or 3D FLAIR sequences, DWI, SWI and post-contrast T1- weighted sequences. Imaging spectrum of findings was documented and MRI patterns were suggested based on these findings.

Results

The study included 27 children; 18 females and 9 males ranging in age from 3 months up to 12 years. Analysis of their attached MRI brain examinations revealed wide spectrum of findings, based on which; three main patterns were identified; normal brain MRI pattern (2 patients), atrophic brain changes pattern (4 patients) and inflammatory brain changes pattern (21 patients). This 3rd group was further subdivided into three sub patterns: dysmyelination/leukodystrophy like pattern noted in five patients, encephalitic/Acute disseminated encephalomyelitis (ADEM) like pattern noted in another five patients and complex pattern which was noted in 11 patients.

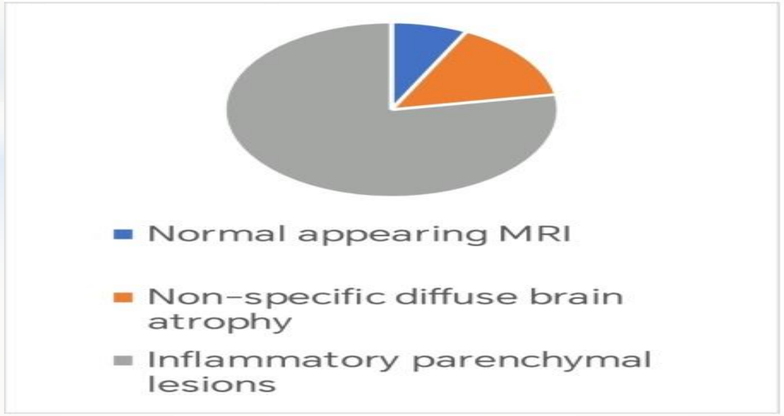

Figure (1): MRI findings in children with CNS-HLH (n = 27)

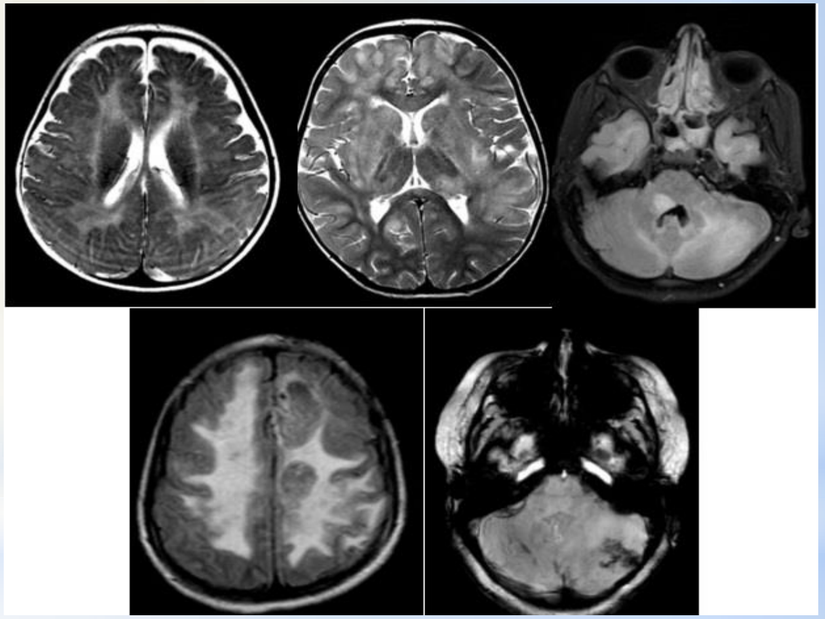

Figure (2): Patterns of MRI in patients with CNS-HLH. A) Axial T2 showing leukodystrophy like pattern. B) Axial T2 showing ADEM like pattern. C&D) Axial FLAIR and F) Axial SWI images showing complex pattern

Conclusion

Haemophagocytic lymphohistiocytosis (HLH) is an underdiagnosed life threatening immunoregulatory pediatric brain disorder. Once HLH diagnosis is suspected based on clinical and laboratory criteria, brain MRI is essential to confirm CNS infiltration. CNS-HLH shows wide spectrum of neuroimaging features ranging from normal brain to inflammatory changes passing by non-specific atrophic brain changes. CNS-HLH imaging patterns may mimic many other pediatric brain diseases including leukodystrophy, encephalitis, ADEM as well as vasculitis which should be taken into consideration in view of associated clinical and laboratory findings suspicious for HLH. Familiarity with such MRI patterns in pediatric CNS-HLH is important and may be life saving for this rare childhood fatal disease.