

monophasic course frequently associated with infections or previous immunization. Regarding pathogenesis, encephalomyelitis is associated with some immunological mechanisms. Post-infection or post-vaccination inflammatory reactions in the perivenular region in the central nervous system (CNS) may be explained by a molecular mimicry mechanism. An antecedent infection was identified in 72–77% of paediatric patients.

We present a case of a 5 year-old girl in follow up for autoimmune thrombocytopenia (AT) and autoimmune haemolytic anaemia (AHA) in steroid treatment, with history of left hemiparesis, language delay, from as a consequence of perinatal suffering, who has developed ADEM.

This patient presented with progressive weakness of limbs, neck pain associated with fever from one day. It was acute in onset and gradually progressive. She became quickly hypotonic and hyporesponsive and had reduced level of consciousness. Within 12 hours of admission, the patient developed sphincter incontinence and dysphagia.

After exclusion of papilloedema, lumbar puncture has been performed. An urgent MRI was performed, which showed multiple subcortical lesions of varying size showing hyperintensities in TR at the bridge and in T2 at the medulla .

Cerebrospinal fluid (CSF) study showed cell count of 40 cells/mm<sup>3</sup> , protein 58 mg/dL, glucose 54 mg/dL. No oligoclonal bands were present in CSF. Blood and CSF cultures were negative like other infectiological analyzes. Also CSF-PCR for the presence of bacteria and virus was negative. Her autoimmune profile with antinuclear antibody was also negative.

The clinical features and the MRI findings were suggestive of ADEM. Partial quadriplegia, and reduction of reflexes, as seen in the myelitic form of ADEM, were present. She had developed ADEM while on the maintenance dose of prednisolone.

After diagnosis, intravenous methylprednisolone was given at 30 mg/kg daily for 5 days. After 48 hours there was a significant improvement in the patient's clinical condition. A regimen of oral steroid was advised after intravenous therapy . The patient responded well to steroid therapy. No residual lesion was found on follow-up.

AT and AHA are relatively uncommon. There are studies postulating the possibility of a combination of several autoimmune diseases. Very few cases have been found with this rare association in the literature. Follow-up of these patients is essential for detecting the development of other autoimmune disease in such cases.

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#### AN UNUSUAL PRESENTATION OF HAEMOPHAGOCYTIC LYMPHOHISTIOCYTOSIS

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**Introduction** We present a case of an unusual clinical presentation of Haemophagocytic Lymphohistiocytosis (HLH). HLH is a rare, life-threatening hyperinflammatory condition resulting from dysregulation of normal innate and adaptive immune responses. The common presenting features of HLH are fever, rash, neurological symptoms, hepatosplenomegaly,

lymphadenopathy, cytopenias, high serum ferritin and liver function abnormalities(2).

**Case** We present a case of HLH in a 5 year old girl presenting to a large regional secondary paediatric unit with unusual clinical features. Although pyrexia is a well-recognised presenting feature of this condition, the predominant features in our case were persistent left-sided pleuritic chest pain with a loud flow murmur and radiological evidence of lower lobe atelectasis with an associated pleural effusion and cardiomegaly. An initial diagnosis of a lower respiratory tract infection on a background history of a yet-to-be-determined cardiac structural defect was made and she was commenced on broad spectrum IV antibiotics and supportive care.

She remained pyrexial and unwell despite 48 hours of IV antibiotics and a repeat chest x-ray revealed radiological deterioration, precipitating a decision to refer her for tertiary paediatric care. Further investigations confirmed a diagnosis of HLH.

**Conclusion** Although rare this serious medical condition can be life threatening and increased awareness is needed, particularly when clinical features at presentation are not characteristic of the condition, to improve early clinical detection, intervention and prognosis.

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#### BLOOD TRANSFUSION IN NICU

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**Introduction** Blood products transfusion is of critical importance to the care of sick and premature infants. Considering the significant drop in hemoglobin level due to the physiological factors after birth till the tenth to twelfth week of life, this study aimed to evaluate the volume of transfused blood in NICU at private hospital, Mashhad, Iran during 6 years.

**Materials and methods** In this cross sectional study, all the infants admitted to NICU of the hospital (2011–2017) were evaluated in terms of the volume of transfused blood. 34% of 24183 patients in this ward were female. 5% of them were preterm. More than 6% weighted less than 2500 gram.

**Results** Of all the admitted patients in this ward, 569 subjects received blood and the related products, most of which 51% (291 neonates) received FFP, and the least 14% (80 neonates) needed PLT. Meanwhile, more than 27% (159 neonates) received PC. It should be noted that most of the neonates (35%) were reported with blood type O<sup>+</sup> and the least (1.5%) with AB<sup>-</sup>.

**Conclusion** Regarding the high volume of transfused blood in NICU, it is essential to focus our attention on the appropriate use of blood and blood products as well as the prevention of transfusion-associated infections.

Although almost more than 70% of NICU patients require less than 20<sup>cc</sup> blood, the adult blood bags are used for this purpose and the fairly large quantities of the blood are discarded. Therefore, it is suggested a special blood bag is designed for neonatal transfusion.