Sultan Qaboos University Med J, August 2023, Vol. 23, Iss. 3, pp. 421–422, Epub. 28 Aug 23
Submitted 30 Jan 23
Revision Req. 19 Mar 23; Revision Recd. 23 Mar 23
Accepted 4 Apr 23
https://doi.org/10.18295/squmj.5.2023.023

INTERESTING MEDICAL IMAGE

Longitudinal Extensive Transverse Myelitis in a Four-Year-Old Boy Liver Transplant Recipient

*Roshan Koul,[1] Shalini Thapar,[2] Rajeev Khanna,[3] Seema Alam[3]

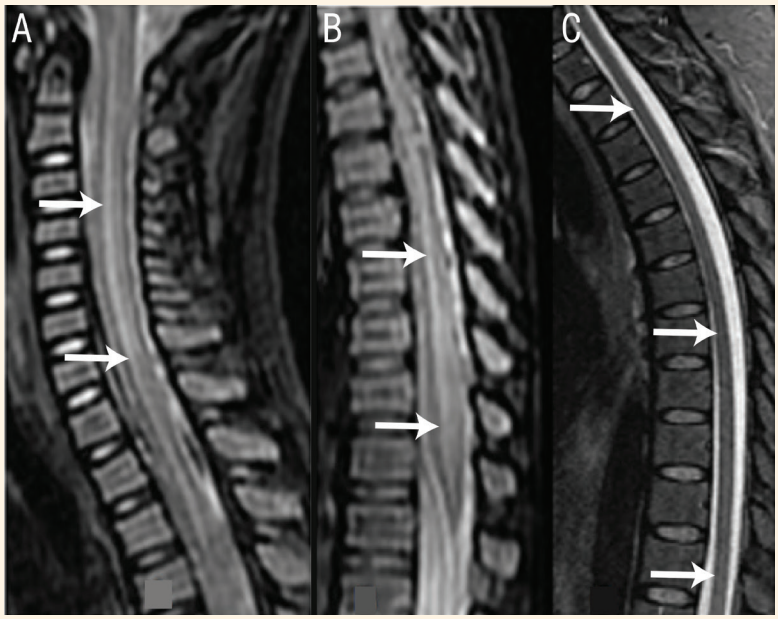

Figure 1: Magnetic resonance imaging short tau inversion recovery (STIR) sagittal image of spine showing (**A**) cervical 1 to dorsal 7 vertebra hyper-intense lesion, (**B**) dorsal 8 to *conus medullaris* lumbar 1 vertebra hyper-intense lesion (the whole spine could not be shown in a single image due to mild scoliosis) and (**C**) a control image showing normal appearance of the spinal cord.

Acute transverse myelitis (ATM) is defined as acute onset quadriplegia or paraplegia with sensory and autonomic involvement progressing in three to four days' time.[1] If the myelitis involves more than three consecutive vertebral segments on magnetic resonance imaging (MRI), it is called longitudinal extensive transverse myelitis (LETM).[1] On Google and PubMed search only, one adult case of transverse myelitis was seen in a liver transplant (LT) recipient.[2] This is the first case of LETM to be reported in a child who had LT.

Four-years-eight-month-old boy diagnosed with biliary atresia during the neonatal period, underwent living liver donor transplantation at the age of one year and five months. He was the third child in the family with no family history of liver disease. He had tubercular lymphadenitis soon after the transplant, which was treated appropriately. He also had prolonged unprovoked seizures in November 2020, which was treated with levetiracetam. He developed an Epstein-Barr virus (EBV) infection and was treated with ganciclovir. He had graft dysfunction due to hepatitis E infection that settled of its own. Two years and ten months after the transplant (2022) he developed a right mandibular mass. Positron emission tomography (PET) computerised tomography (CT) scan showed fluoro-deoxy glucose (FDG) avid irregular mass involving right maxillary sinus, retro-orbital region, and ethmoid sinus with lytic destruction of maxilla. Histopathology revealed Burkett's lymphoma stage IV. Bone marrow and cerebrospinal fluid (CSF) were normal. He was started on rituximab, cyclophosphamide, hydroxydaunorubicin, vincristine, prednisolone (R-CHOP) chemotherapy along with intrathecal methotrexate. After three months, he had received 3 cycles of CHOP with intrathecal methotrexate and 5th pulse of rituximab. He was then admitted with progressive weakness of all four limbs which started in the left arm first and completely paralysed him below the neck in three days' time. His cranial nerves and sensorium were normal. MRI brain was normal and MRI spine revealed almost whole

Departments of [1]Neurology, [2]Radiology and [3]Pediatric Hepatology, Institute of liver and biliary sciences, New Delhi, India
**Corresponding Author's e-mail: koulroshan@gmail.com*

length spinal cord myelitis [Figure 1]. CSF examination was normal. No malignant cells were seen. Viral studies were negative. Aquaporin antibody test was not done as it was not available in the hospital. He was treated with pulse methylprednisolone 30 mg/kg/day for five days followed by oral steroids for four weeks and IVIG 400 mg/kg/day for five days. He started noticing improvement in his weakness after ten days of treatment. Currently, he has grade three power (medical research council grading) in his right upper limb, grade 2–3 in the lower limbs and grade 1 in the left upper limb. Sensory examination was normal all over. He received repeat chemotherapy, including intrathecal methotrexate. There was no worsening of his weakness after intrathecal methotrexate. During the hospital stay he had recurrence of seizures which evolved into status *epilepticus* requiring four anti-epileptic drugs (midazolam, levetiracetam, phenytoin sodium, lacosamide and phenobarbital) to control.

Comment

There are many neurological complications seen in liver transplant recipients.[3] These may be related to the transplant or immunosuppressants used in the treatment.[3] ATM is the rarest of all complications. Only a few reports are available in the literature.[2] Most of the ATM in children are of unknown etiology, believed to be some form of nervous system demyelinating illness in response to varied etiology.[1,4] A thorough workup is required in ATM to rule out infections, autoimmune disorders or infiltrations.[1,4] In a single case of ATM reported in an adult liver transplant recipient, it was correlated to the high levels of tacrolimus.[2] This patient was a 39-year-old male, with chronic liver disease due to Budd-Chiari syndrome. He underwent deceased liver transplant and was put on tacrolimus. He developed weakness of all four limbs on day five post operation. Blood tacrolimus level was normal. MRI brain was normal and MRI spine revealed whole length spinal cord demyelination. No underlying etiology was found on investigation. Immunoglobulins did not improve the weakness. Cyclosporine was added in the treatment and tacrolimus was stopped. This change of immunosuppressant resulted in improvement of his weakness.[2] The current patient had not received tacrolimus and his CSF was normal. Intrathecal methotrexate is widely used and there are a few reports in literature about association of ATM and intrathecal methotrexate.[5] However, the current patient had received three doses previously and the weakness developed after thirteen days of the third dose. Furthermore, he was given a fourth dose without worsening of his neuro status. This observation rules out methotrexate induced ATM. The current patient is the first case of LETM seen in a liver transplant recipient. ATM is uncommon in children.[1,6] LETM presentation in the current patient is possible due to his medical condition, making him more prone to various autoimmune conditions or coincidental. Due to several cofounders in the setting of LT, it is difficult to pinpoint what predisposed this child to LETM. A repeat follow-up MRI spine and clinical monitoring will determine his long-term therapy for the LETM.

AUTHORS' CONTRIBUTION

RoK, SA and RaK managed the patient. RoK provided the diagnosis. ST provided the radiological images. RoK drafted the manuscript and SA edited the manuscript. All authors approved the final version of the manuscript.

References

1. Wolf VL, Lupo PJ, Lotze TE. Pediatric Acute Transverse Myelitis. Overview and Differential diagnosis. J child Neurol 2012; 27:1426–36. https://doi.org/10.1177/0883073812452916.
2. Aksoy F, Dundar HZ, Demir AB, Kiyici M, Kaya E. Myelitis After Liver Transplant: A case Report. Exp Clin Transplant 2021; 1–3. https://doi.org/10.6002/ect.2020.0278.
3. Weiss N, Thabut D. Neurological Complications Occurring After Liver Transplantation: Role of Risk Factors, Hepatic Encephalopathy and Acute (on Chronic) Brain Injury. Liver Transpl 2019; 25:469–87. https://doi.org/10.1002/lt.25420.
4. Scott TE, Frohman EM, De Seze J, Gronseth GS, Weinshenker BG. Evidence based guidline: clinical evaluation and treatment of transverse myelitis: Report of the Therapeutics and Technology Assessment Subcommittee of the American Academy of Neurology. Neurology 2011; 77:2128–34. https://doi.org/10.1212/WNL.0b013e31823dc535.
5. Castillo-Torres SA, Soto-Rincón CA, Villarreal-Montemayor HJ, Chávez-Luévanos B. Case of neuromyelitis optica: bilateral sensorineural hearing loss and transverse myelopathy following intrathecal hemotherapy. BMJ Case Rep 2020; 13:e234076. https://doi.org/10.1136/bcr-2019-234076.
6. Koul R, AlFutaisi A, Mani R, AbdelRahim R, Sankhla DK, AlAzri FM. Longitudinal extensive transverse myelitis in children from Oman. Neurosciences (Riyadh) 2017; 22:127–30. https://doi.org/10.17712/nsj.2017.2.20160352.