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Case Report: Is Intravenous Immunoglobulin Important when Used Together with Pulse Steroid! Complete recovery within 1 Month of Pediatric Patient Diagnosed with Transverse Myelitis

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Abstract

BACKGROUND: Pediatric acute transverse myelitis (ATM) is a potentially devastating condition with variable outcomes and presents significant demands on health and social care resources.

CASE PRESENTATION: The patient in the present study was a 9-year-old boy, unknown to have chronic diseases. He was presented to Maternity and Children Hospital, Tabuk, with a history of acute progressive onset of unsteady gait and imbalance with pictures of encephalopathy. "Change level of consciousness" for >1 week, preceded by fever and upper respiratory tract infection symptoms. Physical findings were very suggestive and consistent with ATM. Diagnosis was confirmed by blood investigations, lumbar puncture analysis, and magnetic resonance imaging (MRI) brain, and spine. The analysis of lumbar puncture (cerebrospinal fluid) showed high white blood cells count and high protein level "more than 500." The findings of MRI brain and spine were consistent with TM. The treatment was done initially with intravenous immunoglobulin (IVIG) by a dose of 2 g/kg divided over 2–4 days; followed by a dose of IV pulse steroid, 30 mg/kg/dose of methylprednisolone once a day for 3 days, with close monitor. The patient had a full physical and radiological recovery within less than one month. In conclusion, IVIG showed a significant role in the management of ATM together with steroid.

CONCLUSION: IVIG showed a significant role in the management of ATM and could be used as a supportive treatment together with IV pulse steroid. IVIG showed a good efficacy with no serious complications.

Introduction

Acute transverse myelitis (ATM) is a focal inflammatory disorder of the spinal cord. The main causes may include systemic autoimmune diseases, localized non-pyogenic infections, multiple sclerosis (MS), post-traumatic and post-infectious events, spinal cord ischemia or hemorrhage, neoplastic and paraneoplastic diseases, and rarely iatrogenic causes [1], [2].

Pediatric ATM is an inflammatory involvement of spinal cord, is a rare demyelinating and immune-mediated disorder of central nervous system (CNS), and contributes to 20% of children experiencing a first acquired demyelinating syndrome (ADS). It may be the first presentation of relapsing ADS such as neuromyelitis optica (NMO) or MS. It can occur, however, as an isolated condition, usually after an infectious disorder. ATM must be differentiated from other presentations of myelopathy (compressive and non-inflammatory myelopathies) [3].

ATM is characterized by abrupt onset of progressive weakness of the limbs, sensory impairment with a sensory level, bladder, and rectal sphincters dysfunction [4].

In a recent center-based analysis of 47 pediatric cases, the age of onset clustered between 0–2 years and 5–17 years. About 47% of the patients had preceding history of febrile illness and 28% had a recent history of vaccination [5].

The tenets of the diagnostic criteria for ATM established by the Transverse Myelitis Consortium Working Group can generally be applied in children; however, a clear sensory level may not be evident in some. magnetic resonance imaging (MRI) lesions are often centrally located with high T2 signal intensity involving gray and neighboring white matter. Longitudinally extensive ATM occurs in the majority. Asymptomatic lesions on brain MRI are seen in more than one-third and predict MS or NMO.

The role of antibodies such as myelin oligodendrocyte glycoprotein in monophasic and relapsing ATM and their significance in therapeutic approaches remain unclear.

ATM is a potentially devastating condition with variable outcome and presents significant cumulative demands on health and social care resources. Children generally have a better outcome than adults, with one-half making a complete recovery by 2 years. There

is a need for standardization of clinical assessment and investigation protocols to enable international collaborative studies to delineate prognostic factors for disability and relapse. There are no robust controlled trials in children or adults to inform optimal treatment of ATM, with one study currently open to recruitment. Thus, the current study provides an overview of current knowledge of clinical features, investigative workup, pathogenesis, and management of ATM, and suggests future directions.

Materials and Methods

Patient and physical examination

Our patient was a 9-year-old boy, medically free "unknown to have chronic diseases," presented to emergency department with a history of acute progressive onset of ataxia (unsteady gait with unclear pattern) and progressive lower limbs weakness with loss of sphincters control over 8 days, and pictures of encephalopathy "change level of consciousness" for <1 week, preceded by a history of fever and headache, no history of seizures (most likely upper respiratory tract infection). Physical findings were very suggestive and consistent with ATM, such as sensory level.

Other systemic review: no cardiac problem and no clear history of frequent choking or chest infection.

Neurological examination: he was noticed to have brisk reflexes all over with power four out of five in both upper and lower limbs with normal cranial nerves examination and sensory level.

Work-up and investigations

Diagnosis was confirmed and established by blood investigations, lumbar puncture analysis, and MRI brain and spine (done with contrast).

Hospital course

In "Maternity and Children Hospital, Tabuk" the patient received a full course of antibiotics and antiviral (full coverage for systemic and CNS infections), which is one of top differential diagnosis to consider in such cases/presentation. Then, he received IV immunoglobulin (IVIG) by a dose of 2 g/kg divided over 2–4 days; followed by IV pulse steroid for 3 days after ID clearance (methylprednisolone 30 mg per kg once a day).

Results

Lumbar puncture (CSF) analysis; showed high WBCs (20 cells/uL) and high protein level "more than 500 mg/L."

MRI brain and spine was done with contrast; findings were consistent with TM (Figure 1 and 2).

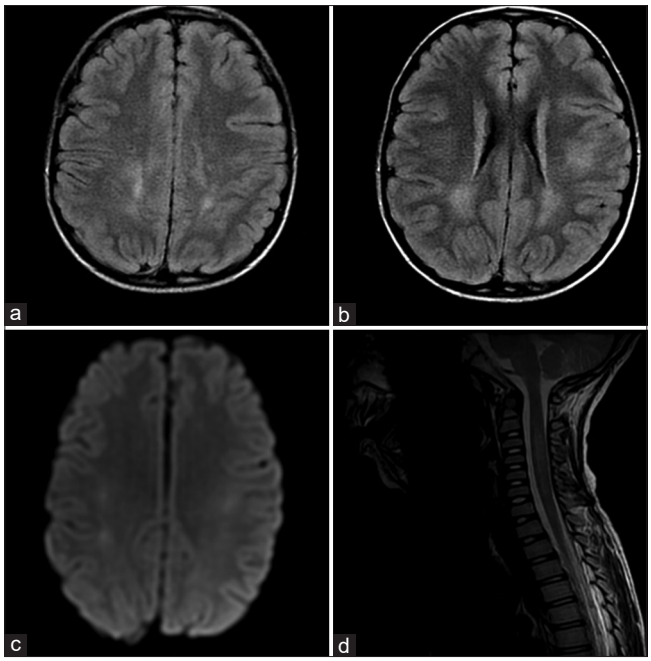

Figure 1: MRI of the brain (a-c) and cervical spine (d); Multiple high FLAIR signal intensities in the deep white matter of the parietal lobes (a and b) with diffusion restriction on the DWI images (c). A long segment (5–6 segments) of high T2 signal intensity occupying more than two-thirds of the cervical cord

The patient had a full physical and radiological recovery within less than one month. He discharged after 2 weeks and few days of admission in good health with complete clinical and radiological recovery with no oral tapering dose.

Discussion

Because of lack of controlled clinical trials, there is no US Food and Drug Administration–approved therapies for ATM. Medications are used based on experience and data from open-label studies and retrospective analyses, primarily from studies involving adults. Data suggest that certain conditions have preferential responses to certain therapeutic interventions.

Intravenous (IV) steroid treatment is often administered for patients with idiopathic ATM. Corticosteroids have various mechanisms of action, including anti-inflammatory properties, immunosuppressive activity, and anti-proliferative actions. In the present study, we used the standard empiric therapy for ATM consists of high-dose corticosteroids. Pediatric patients are usually treated with a 30 mg/kg/dose (maximum 1000 mg) of methylprednisolone intravenously once a day for 3–5 days. Multiple studies have documented the efficacy

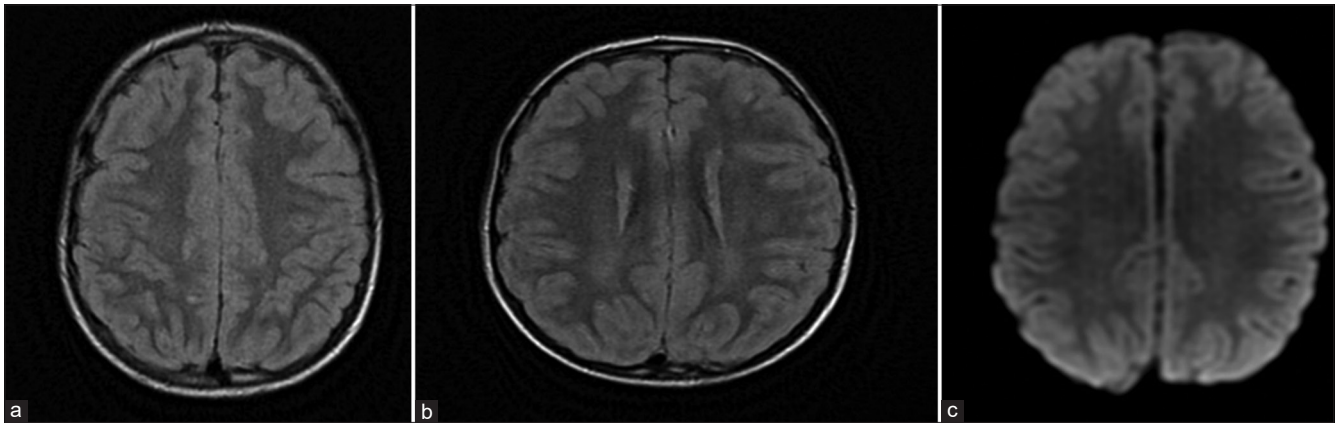

Figure 2: Post-treatment, comparison: almost complete resolution of the high FLAIR signal intensities (a and b) without restriction on the DWI images

and safety of corticosteroids in CNS inflammatory disorders, including ATM. The benefit to patients with ATM was observed in a retrospective study, suggesting better short- and long-term outcomes in patients treated with corticosteroids versus patients who did not receive steroids. Defresne *et al*., [6] reported a favorable effect of high-dose IV methylprednisolone on the proportion of pediatric patients walking independently at 1 month. While, Pidcock *et al*., [5] recently stated that treatment with IV steroids does not improve outcome in children but oral steroids may be associated with improvement in the area of mobility. In spite of these conflicting reports, steroids remain the standard first line intervention for ATM [7].

The results of the current study revealed that IVIG had a significant role in the management of ATM together with steroid. These results were similar to those of De Seze *et al*., [2] who reported in a recent retrospective study on adult cases of idiopathic ATM that two of the four patients treated initially with IV steroids then treated with IVIG showed clinical improvement. In addition, Magraner *et al*., [8] reported that treatment with IVIG is safe and well-tolerated, and it may be used as a treatment alternative for NMO spectrum disorders. Also, Pavlou *et al*., [7] conducted a study on 4-year-old boy who showed clinical and radiological symptoms of myelitis, 10 days after respiratory tract infection, in which a clinical deterioration was observed after administration of corticosteroids, then IVIG was administered and symptoms resolved within 48-h, suggesting an immune-mediated pathogenic mechanism. Contrary, Arango *et al*., [9] reviewed the clinical profile of nine patients in which an investigation of vascular myelopathy was initiated due to worsening of symptoms after treatment with IV steroids, plasma exchange (PLEX) or IVIG, and they concluded that PLEX and IVIG should be avoided during vascular myelopathy.

Anecdotal reports of IVIG 2 gm/kg divided over 2–4 days have not provided conclusive evidence of benefit, but IVIG is often incorporated into the treatment regimen in fulminant disease [10], [11]. A UK randomized controlled trial to determine the benefit of additional treatment with IVIG in adults and children with TM are currently open to recruitment. Further future studies are recommended to test the efficacy of IVIG with or without a high dose of corticosteroids given within a few days after the onset of the disease.

Conclusion

IVIG showed a significant role in the management of ATM and could be used as a supportive treatment together with IV pulse steroid. IVIG showed a good efficacy with no serious complications.

References

1. Krishnan C, Kaplin AI, Deshpande DM, Pardo CA, Kerr DA. Transverse Myelitis: Pathogenesis, Diagnosis and Management. Blue Books of Neurology. Vol. 35. United Kingdom: Butterworth Heinemann; 2010.
2. De Seze J, Stojkovic T, Breteau G, Lucas C, Michon-Pasturel U, Gauvrit JY, *et al*. Acute myelopathies: Clinical, laboratory and outcome profiles in 79 cases. Brain. 2001;124(Pt 8):1509-21. https://doi.org/10.1093/brain/124.8.1509
   PMid: 11459743
3. Tavasoli A, Tabrizi A. Acute transverse myelitis in children, literature review. Iran J Child Neurol. 2018;12(2):7-16.
   PMid:29696041
4. Transverse Myelitis Consortium Group. Proposed diagnostic criteria and nosology of acute transverse myelitis. Neurology. 2002;59:499-505. https://doi.org/10.1212/wnl.59.4.499
   PMid:12236201
5. Pidcock FS, Krishnan C, Crawford TO, Salorio CF, Trovato M, Kerr DA. Acute transverse myelitis in childhood: Center-based analysis of 47 cases. Neurology. 2007;68(18):1474-80. https://doi.org/10.1212/01.wnl.0000260609.11357.6f
   PMid:17470749
6. Defresne P, Hollenberg H, Husson B, Tabarki B, Landrieu P, Huault G, *et al*. Acute transverse myelitis in children: Clinical course and prognostic factors. J Child Neurol. 2003;18(6):401-6. https://doi.org/10.1177/08830738030180060601
   PMid:12886975

7. Pavlou E, Gkampeta A, Kouskouras K, Evangeliou A, Athanasiadou-Piperopoulou F. Idiopathic acute transverse myelitis: Complete recovery after intravenous immunoglobulin. Hippokratia. 2012;16(3):283-5.
PMid:23935301
8. Magraner MJ, Coret F, Casanova B. The effect of intravenous immunoglobulin on neuromyelitis optica. Neurología. 2012;28(2):65-72. https://doi.org/10.1016/j.nrl.2012.03.014
PMid:22841880
9. Arango JJ, Mealy MA, Gailloud P, Villamizar CA. A Note of caution about the diagnosis and treatment of suspected transverse myelitis [TM]. Neurology. 2014;82(10 Suppl):P5. 181. https://doi.org/10.1212/WNL.82.10_supplement.P5.181
10. Absoud M, Greenberg BM, Lim M, Lotze T, Thomas T, Deiva K. Pediatric transverse myelitis. 2016;87(9 Suppl 2):S46-52. https://doi.org/10.1212/WNL.0000000000002820
11. Krishnan C, Kaplin AI, Deshpande DM, Pardo CA, Kerr DA. Transverse myelitis: Pathogenesis, diagnosis and treatment. Front Biosci. 2004;9:1483-99. https://doi.org/10.2741/1351
PMid:14977560