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Neuromyelitis optica associated with systemic autoimmune diseases in children

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Case reports

Neuromyelitis optica (NMO, Devic's disease) is a severe autoimmune disorder predominantly involving optic nerves and spinal cord [1]. Usually isolated, it has been associated with systemic autoimmune diseases in adult patients [2]. We report 2 children with a systemic autoimmune disease who developed NMO. Patients were female; NMO symptoms started at age 8 and 12 years. Patient 1 had a diagnosis of Systemic Lupus Erythematosus (SLE) (fever, cytopenias, mesangeal glomerulonephritis, positive ANA, anti-DNA, anti-Sm). She developed vomiting, tremor, hyperreflexia, paresthesia, neurogenic bladder and progressive vision loss 2 years after SLE onset. Neuroradiological investigations disclosed longitudinally extensive transverse myelitis and bilateral optic nerve involvement. Patient 2 had recurrent parotitis for 1 year before she developed vision loss, papilitis, dystonia and paresthesia. She exhibited positive ANA, anti-Ro and anti-La, and objective eye dryness. MRI evidenced lesions in brainstem and spinal cord, and evoked potentials revealed optic nerve involvement. A diagnosis of Sjögren's Syndrome (SS) associated with NMO was made. NMO-Ig was detected in both patients' sera. Intravenous and p.o. high dose steroids, a 6-month course of monthly I.V. cyclophosphamide (up to 1 g/m^2/dose), followed by azathioprine (2–3 mg/Kg/day) as maintenance therapy, were used in both (plasmapheresis in one). Visual, motor and sensitive symptoms dramatically improved. Mild relapses occurred in both children; they were successfully treated with steroids. Patients are currently well, with improved vision and residual lesions in MRI.

Conclusion

NMO can occur in the setting SLE or SS in children. Intensive immunosuppressive therapy may induce remission and prevent visual loss.

References

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