Luaran Miastenia Gravis Okular pada Remaja dengan Efek Samping Steroid dan Gizi Kurang

Savira*, Agung Triono**, Retno Palupi Baroto**
* Residen Ilmu Kesehatan Anak, Fakultas Kedokteran, Kesehatan Masyarakat, dan Keperawatan Universitas Gadjah Mada, Yogyakarta, Indonesia
**Departemen Ilmu Kesehatan Anak, Fakultas Kedokteran, Kesehatan Masyarakat, dan Keperawatan Universitas Gadjah Mada, Yogyakarta, Indonesia

INTISARI

Pendahuluan: Miastenia Gravis (MG) merupakan kelainan autoimun akibat adanya gangguan transmisi sinaps pada tautan neuromuscular yang menyebabkan kegagalan kontraksi otot. Manifestasi MG berupa kelemahan otot yang bersifat progresif setelah beraktivitas. Gejala MG yang terbatas pada otot okular disebut sebagai MG okular. Dalam dua tahun pertama, 70-85% MG okular pada anak dapat berkembang menjadi MG umum. Tujuan laporan kasus ini untuk membahas tata laksana pasien anak dengan MG okular serta mencegah kejadian MG umum.

Presentasi Kasus: Anak laki-laki berusia 10 tahun terdiagnosis MG okular sejak usia 8 tahun. Telah dilakukan pemeriksaan Repetitive Nerve Stimulatory (RNS) dengan hasil adanya decrement lebih dari 10% pasca latihan, dengan kesimpulan mengarah kelainan MG. Hasil MSCT dan MRI kepala tidak menunjukkan adanya kelainan intrakranial. Hasil pemeriksaan OMD dan foto polos dada menunjukkan tidak ada gangguan menelan dan tak tampak gambaran timoma. Pasien telah mendapat terapi piridostigmin dan steroid, tetapi didapatkan adanya efek samping steroid berupa glaukoma. Gejala ptosis menetap, sehingga dilakukan penghentian dosis steroid dan diberikan terapi azathioprine 2.5mg/kg/hari. Lima bulan pasca pengobatan dengan azathioprine didapatkan perbaikan klinis berupa gejala diplopia membaik, perbaikan paresis nervus kranialis III, IV, dan VI, akan tetapi gejala ptosis menetap. Tidak didapatkan adanya efek samping obat baik secara klinis maupun abnormalitas hasil laborat.

Kesimpulan: Pada pasien anak dengan MG okular yang tidak membaik dengan terapi steroid, dapat dipertimbangkan pemberian azathioprine sebagai agen imunosupresan lain. Pemantauan klinis dan tata laksana adekuat dapat memperbaiki gejala klinis dan mencegah kejadian MG umum.

Kata kunci: okular; miastenia gravis; remaja; *azathioprine*

Outcome of Ocular Juvenille Myasthenia Gravis with Side Effect of Steroid and Moderate Acute Malnutrition

Savira*, Agung Triono**, Retno Palupi Baroto**
*Pediatric Resident, Department of Child Health, Faculty of Medicine, Public Health, and Nursing, Universitas Gadjah Mada, Yogyakarta, Indonesia
**Department of Child Health, Faculty of Medicine, Public Health, and Nursing, Universitas Gadjah Mada, Yogyakarta, Indonesia

ABSTRACT

Introduction: Myasthenia Gravis (MG) is an autoimmune disorder caused by disruption of synaptic transmission in the neuromuscular link which causes failure of muscle contraction. The manifestation of MG is progressive muscle weakness after activity. MG symptoms that are limited to the ocular muscles are referred to as ocular MG. In the first two years, 70-85% of ocular MG in children can progress to generalized MG. The aim of this case report is to discuss the management of pediatric patients with ocular MG and prevent the occurrence of common MG.

Case Presentation: A 10 year old boy was diagnosed with ocular MG since the age of 8 years. A Repetitive Nerve Stimulatory (RNS) examination has been carried out with results of decrement of more than 10% after exercise, with the conclusion pointing to MG abnormalities. The results of MSCT and MRI of the head did not show any intracranial abnormalities. The results of the OMD examination and plain chest x-ray showed that there were no swallowing disorders and no thymoma was visible. The patient had received pyridostigmine and steroid therapy, but found the side effect of steroids in the form of glaucoma. Symptoms of ptosis persisted, so the steroid dose was discontinued and azathioprine therapy 2.5 mg/kg/day was given. Five months after treatment with azathioprine, there was clinical improvement in the form of improved symptoms of diplopia, improvement in paresis of cranial nerves III, IV, and VI, but the symptoms of ptosis persisted. There were no side effects of the drug either clinically or laboratory results abnormalities.

Conclusion: In pediatric patients with ocular MG who do not improve with steroid therapy, azathioprine can be considered as another immunosuppressant agent. Clinical monitoring and adequate management can improve clinical symptoms and prevent the occurrence of common MG.

Keywords: ocular; myasthenia gravis; juvenille; azathioprine