Laporan Kasus
Luaran Anak Ensefalitis Autoimun Anti-NMDAR (N-Methyl-D-Aspartate Receptor)

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Abstrak: Ensefalitis anti-N-metil-D-aspartat reseptor (NMDAR) adalah kelainan autoimun dengan gejala defisit neurologis dan psikosis akut. Kami melaporkan seorang anak perempuan berusia 8 tahun dengan gejala demam, kejang pada wajah dan ekstremitas atas, defisit memori, agitasi, bicara dan kesulitan menelan. Tusukan lumbal, HSV 1 serologis, dan HSV 2 normal. Pasien dirawat di PICU selama sebulan oleh dan didiagnosis sebagai ensefalitis anti-NMDAR. Pasien mendapatkan Intravena Immunoglobulin (IVIg) 0,4 gr/kg/hari selama 5 hari dan diikuti dengan metilprednisolon 30mg /kg/hari. CT scan menunjukkan adanya ventrikulomegali sininstra. MRI dan EEG dilakukan setelah 6 bulan terapi dengan hasil yang konsisten dan epileptiform difus abnormal. Hasil BERA normal. Evaluasi EEG menunjukkan ritme dasar yang lebih baik. Pasien menjalani protokol sitostatika selama 30 bulan terdiri dari metilprednisolon intravena 30mg/kg/hari setiap bulan diikuti dengan pemberian oral harian 0,5-2 mg /kg/hari dengan cyclophosphamid 500-1000mg /m^2 setiap bulan. Pasien merespons imunoterapi dengan baik tanpa keterlibatan kognitif. Pemeriksaan hasil kecerdasan menunjukkan IQ 93 (normal) dengan peningkatan dari kualitas hidup. Diagnosis definitif serum Anti-NMDAR menunjukkan hasil positif yang lemah setelah 29 bulan terapi.
Kesimpulan: Ensefalitis anti-NMDAR bermanifestasi sebagai psikotik akut dengan kejang. Diagnosis yang terlambat mengakibatkan keterlambatan perawatan. Namun, meskipun administrasi protokol terapi terlambat, perbaikan yang signifikan dicatat setelah 30 bulan pengobatan metilprednisolon, IVIg dan siklofosfamid.
Kata kunci: reseptor NMDA, ensefalitis, autoimun, psikosis akut, anak-anak, kejang

Abstract: Anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis is an autoimmune disorder with symptoms of neurological deficits and acute psychosis. We reported an 8-year-old girl with symtoms of fever, seizure of facial and upper extremities, memory deficits, agitation, speech and swallowing difficulties. Lumbar puncture, serological HSV 1 and HSV 2 were normal. The patient was admitted to PICU for a month by and diagnosed as anti-NMDAR encephalitis. The treatment were Intravenous Immunoglobulin (IVIg) 0.4 gr/kg/day for 5 days and followed by methylprednisolone 30mg/kg/day. CT scan shows the presence of sininstra ventriculomegaly. MRI and EEG were performed after 6 months of therapy with consistent results and abnormal diffuse epileptiform. BERA result that time was normal. Evaluation of EEG showed better basic rhythm. Patients who undergo the sitostatics protocol for 30 months consist of intravenous methylprednisolone 30mg/kg/day every month followed by daily oral oral 0.5-2 mg/kg/day with cyclophosphamid 500-1000mg/m^2 every month. Patients responded well to immunotherapy with no cognitive involment. Examination of intelligence quotient showed 93 (normal) with improvement from quality of life. A definitive diagnosis of serum Anti-NMDAR showed a weak positive result after 29 months of therapy.
Conclusion: Anti-NMDAR encephalitis manifests as acute psychotic with seizure. Late in diagnosis resulting in the late of treatment. However, despite of late administration of therapeutic protocol, significant improvement was noted after 30 months treatment of metilprednisolon, IVIg and cyclophosphamide.
Keywords: NMDA receptor, encephalitis, autoimmune, acute psychosis, children, seizure