Neuropsychiatric Systemic Lupus Erythematosus (NPSLE)

Case presentations and topic discussion

The Rheumatology Unit UMMC experience
References


CNS Lupus – Current scenario in UMMC

- From the beginning of 2005 an average of 1 admission a month (new case).
- Varied presentations closely following the revised ACR criteria (19 NPSLE definitions)
- Problems with establishing a concrete diagnosis and we lack a clear treatment protocol.
CNS Lupus

- Nervous system manifestations are present in up to 70% of patients with SLE.
- There are 19 definitions which are components of NPSLE. The earlier classifications only recognized 2 clinical entities – seizures and psychoses
NPSLE based on the revised ACR criteria

- Acute confusional state
- Anxiety disorders
- Cognitive function impairment
- Affective disorders
- Psychoses
- Cranial nerve disorders
- Mononeuropathy
- Plexus neuropathy
- Vegetative neuropathy
- Myasthenia
- AIDP
- Polyneuropathy
NPSLE component entities

- Epileptic attacks
- Headaches and migraines
- Cerebrovascular diseases
- Demyelinating syndromes
- Aseptic meningitis
- Chorea
- Myelopathy
CNS Lupus – problems in diagnosis

- Subtle presentations – wouldn’t I be depressed or anxious (or both) if I was diagnosed with SLE?
- Effect of corticosteroids
- Other differential diagnoses?
  Infections and metabolic diseases
- Access to imaging facilities
NPSLE specific antibodies

- Anti ribosomal P antibody
- Anti neuronal antibodies
Imaging

- MRI by far the most superior
- SPECT scans have also been found to be useful
- In the UMMC experience the MRI has shown to be of great diagnostic value
Case 1 – 14 years old Indian girl who presented to Kuantan with dense right sided hemiplegia who showed remarkable recovery with intravenous Methyl Prednisolone.

Low complements, anti dsDNA 276 iu/mL, Ig ACL 4 units, LA negative
12 years old Chinese girl presented with seizures while under treatment for class IV lupus nephritis.

- Low complements anti dsDNA 167 iu/mL, IgG ACL 4 units, LA negative.
- Cases presented today had CT scans with contrast which were normal but with low complements and anti ds DNA in the thousands.
NPSLE - CSF

- Varied findings among samples from our cases, an indicator which we use is the CSF proteins (tend to be higher than 0.45)
- More useful to rule out infections. **Always** send for TB culture.
Therapeutic approach in CNS lupus – mild disease

- Symptomatic therapy
- Analgesics
- Anxiolytics
- Antidepressants
- Tricyclics
- Fluoxetine
- Anti convulsants
- Anti psychotics
- Low dose corticosteroids
Therapeutic approach in CNS Lupus – severe CNS disease: diffuse/nonthrombotic disease

- Acute treatment
- High dose corticosteroids
- Iv pulse methylprednisolone
- Iv pulse cyclophosphamide
- Plasmapharesis
- Iv immunoglobulins
- Intrathecal methotrexate
- Azathioprine
- Mycophenolate mofetil
Therapeutic approach in CNS Lupus – severe CNS disease: diffuse/nonthrombotic disease

- Chronic treatment
- Taper corticosteroids
- Iv pulse cyclophosphamide
- Methotrexate
- Azathioprine
- Mycophenolate mofetil
Current practice at UMMC

- Intravenous methylprednisolone 500mg daily for 3 days followed by one of 2 cyclophosphamide regimes
  - 1) monthly courses of 1.0 grams/BSA for 6 months followed by 3 monthly courses.
  - 2) 2 weekly courses of 500mg total of 6 doses (3 grams in total)
Current practice at UMMC

- Symptomatic treatment is accompanied by intravenous methylprednisolone and high dose 1mg/kg of oral prednisolone daily which is tapered. Usually a steroid sparing agent such as azathioprine is added.
- Hydroxychloroquine
- Role for cyclophosphamide in such patients? At which doses – 1 or 2?