

PRIMARY NEUROPSYCHIATRIC PRESENTATION IN SYSTEMIC LUPUS ERYTHEMATOSUS

Background:

Neuropsychiatric manifestations of systemic lupus erythematosus occur in up to 75% of patients. However, it is much less frequent that such manifestations occur as an initial clinical event.

Aim:

We report a case of CNS lupus, presenting primarily with neuropsychiatric symptoms.

Patients and methods:

An African 7-year-old girl was admitted to our hospital with a 4-day history of ataxia, diplopia and morning vomiting. She also had complaints of severe headache, cyclic vomiting and psychiatric symptoms - auditory hallucination, irritable mood and aggressive behaviour - beginning 1 year earlier.

She had no fever or meningeal signs and she presented with a broad-based, unsteady gait, bilateral dysmetria, horizontal nystagmus and hyporeflexia of the lower members. Laboratory studies showed a normocytic, normochromic anemia, a normal leucocyte count, a raised erythrocyte sedimentation rate with a normal C reactive protein. The brain MRI showed multiple cortico-subcortical lesions in both cerebral hemispheres with increased signal intensity in long TR sequences, highly suggestive of encephalitis, considered as "possibly herpetic".

Despite sterile CSF with normal cytochemical markers, she began treatment with acyclovir, ceftriaxone, tuberculostatics and dexamethasone. CSF bacteriologic, mycologic and mycobacteriologic cultures and PCR for multiple viral, bacterial and fungal agents were negative. Immunoelectrophoresis showed intrathecal synthesis of IgM and oligoclonal IgG bands.

Immunological markers revealed a high-titre of ANA (1:5120) and positive anti-dsDNA antibodies.

Results:

Immunosuppressive treatment with methylprednisolone, cyclophosphamide and oral prednisolone was started, with marked improvement of the clinical condition and resolution of nearly all cerebral lesions on MRI.

Conclusions:

In an unusual encephalitis presentation, a wide range of differential diagnosis has to be considered. Primary neuropsychiatric Lupus may present very difficult diagnostic and therapeutic challenges.

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