The Radiological Case of the month

Acute Disseminated Encephalomyelitis (ADEM) In A Four Year Sudanese Girl.

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A 4 year old girl brought to the hospital suffering from fever, headache, convulsions & behavioral changes. On examination she was febrile, pale, with choreoathetoid movements & stupor that progressed rapidly to a state of coma. There were positive signs of meningeal irritation. GCS was 7 at the start. There was head lag & Rt. 6th cranial nerve palsy. She was spastic all over with exaggerated reflexes. Got upgoing planter reflex. Later she developed typical Decorticate position.

CBC, Blood grouping & BF for malaria random blood sugar, renal profile & ABG. MRI was done few days later, while on treatment, which is shown above were also done.

Management was as follows: IVFs, NG tube, valium, IV antibiotics in form of ceftriaxone, Ampiclox & Gentamycin. Acyclovir, dexamethazone & metronidazole were also added.

She responded to our management, but with the following sequelle: Aphasia, epilepsy & mental retardation. So an antiepileptic drug and physiotherapy were started.

ADEM is an acute demyelinating disease, of autoimmune etiology, most commonly affects children. It is a white matter inflammatory disease. It usually follows a viral infection, a nonspecific upper respiratory infection or vaccinations. ADEM is predominantly a disease of children and in particular infants. Clinically there is fever, headache, stiff neck with multifocal deficits (Ataxia, hemiplegia & visual loss). The child may get fits, stupor or coma.
ADEM ultimately has a better prognosis. Most cases have favourable outcome, but few are severe or fatal. Diagnosis carries important therapeutic and prognostic implications. CSF shows: An increase in WBC & protein level. Rarely there is what is called ADEM-like presentation. Multiple sclerosis is the main differential diagnosis.

MRI is the neuroimaging study of choice for establishing the diagnosis and for following the course of the disease. The MRI shows: Multiple bilateral hyperintense lesions & perilesional edema. CT scan is relatively insensitive. The lesions regress with successful treatment. Treatment starts with IV methylprednisolone for three days followed by oral steroids for few weeks. Infusions of immunoglobulins can be given.

A study done in North Indian children stated that: The presentation of paediatric ADEM in developing countries is similar to that in developed countries. In spite of an aggressive presentation, most children respond well to corticosteroids. MRI lesions disappear or are significantly reduced at six months in the majority of cases.

References:
(2) Bennetto L, Scolding N. Inflammatory post-infectious encephalomyelitis. J Neurol Neurosurg Psychiatry. 2004; 75: 2228-.