Acute Demyelinating Encephalomyelitis Presenting as PUO

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Introduction

Acute Disseminated Encephalomyelitis (ADEM) is an immune-mediated inflammatory disorder of the central nervous system. It usually follows a viral prodrome. Symptomatology includes predominantly long tract signs with or without involvement of optic nerve tract which is manifested as alteration in visual acuity and altered Visual evoked potential. Diagnosis is usually confirmed by MRI Brain or spinal cord which typically shows changes of demyelination. Rarely it can only present as Pyrexia of Unknown Origin with neurological symptoms and signs appearing later during the course of illness [1]. We report 3 children who presented as PUO and eventually were diagnosed as ADEM.

Case 1

5 year old boy presented with prolonged fever with no focus of infection with raised inflammatory markers. During the 2nd week of illness he developed an ataxic gait and was found to have exaggerated DTRs. MRI spine showed features of demyelination and he improved with MethylPrednisolone.

Case 2

9 year old girl presented with prolonged fever with no focus of infection and her cultures were sterile. However she had high inflammatory markers and hence 2nd line investigations were done to rule out Kawasaki disease and autoimmune disease which were negative. During the course of illness she developed visual blurring which was evidenced by decreased visual acuity and impaired visual evoked potential. Her MRI brain was suggestive of demyelination changes and she improved with Pulse MethylPrednisolone.

Case 3

5 year old girl resident of Japan presented with prolonged fever with no focus of infection. During the 2nd week of illness she became encephalopathic and developed long tract signs. Her MRI brain was suggestive of ADEM and she too improved with Pulse MethylPrednisolone.

We suggest that ADEM should be considered in the differential diagnosis of pyrexia of unknown origin in children, even in the absence of any neurological manifestations, as early treatment can be associated with clinical improvement and may prevent serious complications. A successful diagnosis of the underlying disease requires an intensive and rational diagnostic evaluation of the wide spectrum of possible etiologies of PUO. Periodic clinical examination will give us valuable information judicious use of second line investigations in a child with PUO and close follow up of these patients is important [2].

References: