A RARE CASE OF ACUTE INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY AS FIRST PRESENTATION IN A HIV INFECTED CHILD - A CASE REPORT
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Abstract: Acute inflammatory demyelinating polyradiculoneuropathies (AIDP) occur with increased frequency in people with HIV, particularly in the early stages of infection, and probably represent an autoimmune phenomena. The clinical and electrophysiological features, as well as the response to immunomodulating therapies are indistinguishable from those seen in non-HIV associated AIDP. However the classic finding of albuminocytological dissociation, where cerebrospinal fluid (CSF) protein is elevated but the CSF is acellular, may be absent, because typically in HIV-associated AIDP, there is a lymphocytic pleocytosis in addition to elevated CSF protein.

Keyword: AIDP, HIV

CASE REPORT:

Child having asymmetrical weakness of all four limbs
2.5 year old 1st born male child of nonconsanguinous marriage presented to us with subacute onset of weakness of both lower limbs followed by weakness of both upper limbs over a period of 10 days. He had history of trunk muscle weakness. There was no history suggestive of cranial nerve dysfunction, bladder & bowel involvement or sensory disturbances. The child had no history of trauma /altered level of consciousness /seizures /recent exanthematos fever / recent vaccination or dogbite. The child was breast fed upto two years of age. There was no history of developmental delay.

All the nerves were unstimulatable
On examination, child was conscious, afebrile. His milestones were normal. He showed no obvious facial dysmorphism. His vitals were normal. CVS and RS were clinically normal. There was no organomegaly. Neurological examination revealed normal cranial nerves. Motor examination revealed generalized hypotonia. Muscle bulk was normal. Power was 2/5 in all 4 limbs. There was areflexia of all 4 limbs and bilateral plantar reflex was absent. Spine & cranium were normal. There were no meningeal signs. MRI study of the brain and spinal cord was normal. On nerve conduction study, all nerves were unstimulatable. CSF analysis showed protein - 62, sugar-75, cells – 10 lymphocytes; typical albuminocytological dissociation was absent. Further investigation revealed child was HIV positive and CD4 count was 280/μl. After repeated history taking father was found to be HIV positive. Further testing of mother showed HIV positive.

Child’s muscle power improved after IV Ig
Child was treated with IV Immunoglobulin 400 mg/kg X 5 days and anti-retroviral therapy was started. Child was given physiotherapy. Power improved to 4/5.

DISCUSSION:
HIV infection is known to cause a variety of neurologic conditions. The case underlines the importance of appropriate testing for HIV infection in patients with GBS. In addition to its tendency to invade immune cells, HIV has a propensity to infect nervous tissue. This "neurotropism" occasionally results in clinically apparent disease early in the course of HIV infection, often during the time of the acute retroviral syndrome. Some of the neurologic manifestations of acute HIV infection include aseptic meningitis, encephalitis, myelopathy, and various peripheral neuropathies. Rarely, acute inflammatory demyelinating polyradiculoneuropathy, or Guillain-Barré syndrome (GBS), has been associated with HIV seroconversion (1). GBS is a relatively uncommon neurologic disease with an annual incidence estimated at 1.7 per 100,000.
Our case highlights the importance of screening previously healthy persons with GBS for HIV infection. It is important for physicians to be aware of this relationship and to obtain a detailed history at the time of presentation, focusing on potential risk factors for HIV infection. The characteristic CSF finding in GBS is one of albuminocytologic dissociation—namely, an elevated CSF protein level with a normal WBC count. Therefore, pleocytosis in a patient whose clinical history is consistent with GBS should prompt an evaluation for HIV infection. Pleocytosis is commonly seen in acute HIV infection, as well as in advanced HIV infection (2,4). A nonspecific infectious syndrome precedes the onset of GBS in a majority of patients. The associated pathogens include C. jejuni, Mycoplasma, Epstein-Barr virus, cytomegalovirus, Borrelia, and HIV (10). There have been 4 reported cases of GBS occurring during acute HIV seroconversion in adults (6).

Various mechanisms have been proposed to explain the relationship between HIV and GBS. One theory is that HIV infection causes a dysfunction in immune regulation resulting in an autoimmune attack on neural myelin sheaths. Additional neural damage from HIV-1 infection may be caused by neurotoxins produced by infected or activated macrophages or monocytes or by toxic products of HIV-1 itself. Direct invasion of nerves by HIV has also been postulated (4). Despite earlier evidence that HIV can directly infect nervous tissue, attempts to isolate the virus from peripheral nerves in GBS patients have had inconsistent results. Recently, there has been some success in identifying HIV-1 DNA in neurons in the CNS using laser capture microdissection (7). While GBS is primarily a disease of peripheral nerves, evidence that HIV has the capacity to infect neurons suggests that direct infection could play a role in GBS.

The clinical course of HIV-related GBS and non-HIV-related GBS are similar in terms of clinical manifestation and immune therapy. Immunoglobulin and plasma exchange are equally effective. Our patient's clinical improvement following treatment with high-dose IVIg supports an earlier report of 2 HIV-positive patients who improved after IVIg therapy (8). Furthermore, these patients may benefit from antiretroviral therapy at the time of seroconversion. This case highlights the importance of having a high degree of suspicion for HIV infection in patients who present with GBS. It should be noted that the majority of HIV-positive patients presenting with GBS already have established retroviral infection. Nonetheless, physicians should be aware of this less common association between GBS and acute HIV seroconversion (1).

HIV testing should be conducted in patients who have risk factors for HIV infection, evidence of generalized lymphadenopathy on physical examination, or leukocytosis on CSF analysis. Test results may be falsely negative or indeterminate early in the course of HIV infection and, therefore, should be repeated in these patients (2). Alternatively, a plasma HIV load or p24 antigen level can be obtained.

REFERENCES:
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