

HIV ENCEPHALOPATHY IN A CHILD: A RARE PRESENTATION

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ABSTRACT

Human Immunodeficiency virus encephalopathy is a rare presenting feature in children. We had a 7 year female child of HIV positive parent, who presented with fever, progressive decline in neurological function and altered sensorium for 15 days. On examination, she was febrile, unconscious (GCS 4/15) with decerebrate posturing, symmetrical exaggerated deep tendon reflexes, and bilateral extensor plantar. Her ELISA for HIV was positive and CD4+ count was 89 cells/ μ l. CSF cytochemical analyses was normal with negative India ink study for cryptococcus. Cranial CT showed brain atrophy in serial scans. A diagnosis of progressive HIV encephalopathy was made and antibacterials, antifungals and HAART regimen was started but child succumbed to illness. Progressive encephalopathy in childhood may be due to HIV infection.

KEY WORDS: HIV, Encephalopathy, Infection

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INTRODUCTION

A wide range of neurologic manifestations related to infection with human immunodeficiency virus have been reported in children with perinatally acquired disease.^{1,2} A progressive encephalopathy with cognitive, behavioral, and motor manifestations has been described, as has been a more static form of neurologic impairment. In the majority of the children with neurologic dysfunction, no identifiable process or specific pathogen other than HIV can be found. Compelling evidence exists that the central nervous system (CNS) can be directly infected by HIV, and neuropathologic changes may be associated with this infection.³ It has also been shown that antiretroviral therapy can improve the neuropsychologic functioning of children whose CNS has been affected.⁴

CASE REPORT

A 7 year female was brought by her grandparents to pediatric emergency with fever for 1.5 months, progressive decline in neurological function and unconsciousness for 15 days. Child was treated for 15 days outside before admission in our hospital. Fever was high grade (103°F), continuous, without chills and rigors. Neurological decline in the form of not interested in surroundings, reduced activity, irrelevant talk after which child lapsed into altered sensorium. There was no history of vomiting, headache, seizure, cough, skin rashes and jaundice. Both mother and father were HIV positive and mother expired 3 years back.

On examination, she was unconscious (GCS 4/15), febrile with vitals, respiratory rate:48/min, Pulse:130/min and BP: 96/70 mm of Hg. On neurological examination, she had symmetrical exaggerated deep tendon reflexes in all limbs, bilateral extensor plantar response and decerebrate posturing. The examinations of chest cardiovascular system and abdomen were clinically normal.

Her hemogram showed Hb: 9.5 gm%, TLC:7700 with differential of 78% polymorphs, 16% lymphocytes and platelets 86,000/ cu mm. CSF routine cytochemical study was normal and india ink study for cryptococcus was negative. ELISA using 3 different kits for HIV was positive. CD4+ cell count was 89 cells/ μ l. CT scan of brain done 1 month earlier and at admission showed diffuse brain atrophy without any meningeal enhancement or parenchymal lesions (Figure 1 & 2). Tuberculin skin test was negative and chest radiograph was normal. Arterial blood gases and serum electrolytes were normal. Urine routine microscopy was normal. Thus a diagnosis of progressive HIV encephalopathy was made in this case based on clinical laboratory and imaging study.

Patient was treated conservatively with oxygen, intravenous fluid and ceftriaxone awaiting report of CSF analysis. As

child's general condition remained same with fever persisting, intravenous fluconazole was added after 48 hours. HAART was added on 5th day of admission with stavudine, lamivudine and nevirapine. In spite of all the above measures, child succumbed to illness on 9th day of admission.

DISCUSSION

The exact incidence of CNS involvement in HIV infection is not known, it is thought to occur in most HIV- infected children⁵ and its incidence is three times more in children than that of adults⁶. HIV encephalopathy is common in HIV- infected children who present in early infancy and have rapid downhill course⁵. The risk of HIV encephalopathy is also correlated directly with the severity of HIV-related symptoms, depression of CD4+ counts and p24 antigen levels in the mother⁵. The cumulative incidence of HIV-1 related encephalopathy at 7 years post- infection was reported to be 16% in children and the incidence of encephalopathy was 9.9% in the first year of life, 4.2% in the second year and less than 1% per year thereafter⁶. This underscores the fact that HIV encephalopathy can occur very early in the course of HIV infection and that 88.1% of children who develop encephalopathy do so within first two years of life.

The virus probably enters the CNS through infected macrophages⁵. The neurological manifestations may be effected through the direct effects of the virus or through cells of macrophage lineage and toxic cytokines⁵. Developmental delay (motor and cognitive) is a common feature. The encephalopathy may manifest suddenly as a loss of attained milestones or failure to attain new milestones. This may be interspersed with periods of relative stability or rapid deterioration. Other neurological manifestations include pyramidal tract involvement (spastic paraparesis, hypertonicity, hyperreflexia), static encephalopathy with developmental delay without loss of previously attained milestones, progressive encephalopathy with progressive deterioration of milestones and skills, acquired microcephaly, seizures, behavioral decline and cerebral arteriopathy with intracerebral aneurysms.^{5,6}

The diagnosis HIV encephalopathy is based on CDC revised classification system⁶ i.e. presence of ≥ 1 progressive neurological findings for at least two months (in the absence of other identifiable causes) from amongst the following:

1. Failure to attain or loss of milestones or of intellectual ability, verified by standard, developmental scale or neuropsychological tests;
2. Impaired brain growth or acquired microcephaly as demonstrated by head circumference measurements or brain atrophy on CT scan/MRI (serial imaging is required for children less than two years of age);

- Acquired symmetric motor deficits with two or more of the following: paresis, pathologic reflexes, ataxia or gait disturbance.

A simplified case definition of HIV encephalopathy as suggested by Newton et al, 2006 include; children who is HIV positive, lack of growth in head circumference at least 3 months apart, altered neurodevelopmental milestones and lack of acquisition of skills, diffuse symmetrical hyperreflexia, and normal CSF study to exclude CNS infections.

Over half of the patients succumb to the illness within 3 years of diagnosis and the median survival rate in patients with HIV-encephalopathy is about 11 months from the diagnosis⁵. Before treatment, progressive HIV encephalopathy (PHE) was reported to occur in 35% to 50% of children with AIDS and 21% to 35% infected with HIV. After the introduction of anti-retroviral therapy, the incidence of PHE decreased to <2%. PHE in the post HAART era is thus an infrequent and reversible complication of HIV infection that responds to effective anti-retroviral therapy and that may relapse if viral control is lost. Evolution of PHE also has changed since the introduction of anti-retroviral therapy⁷. Viral Load (either baseline or at time of diagnosis) is predictive of PHE but CD4 count did not⁸.

Our patient succumbed after 5 days of starting HAART, which emphasize that unexplained progressive encephalopathy warrants evaluation for HIV at the earliest. Maternal death gave clue in our case, otherwise a child presenting with fever followed by rapidly progressive encephalopathy may have difficulty in making diagnosis.

Figure 1: Cranial tomography showing cerebral atrophy 1 month prior to admission

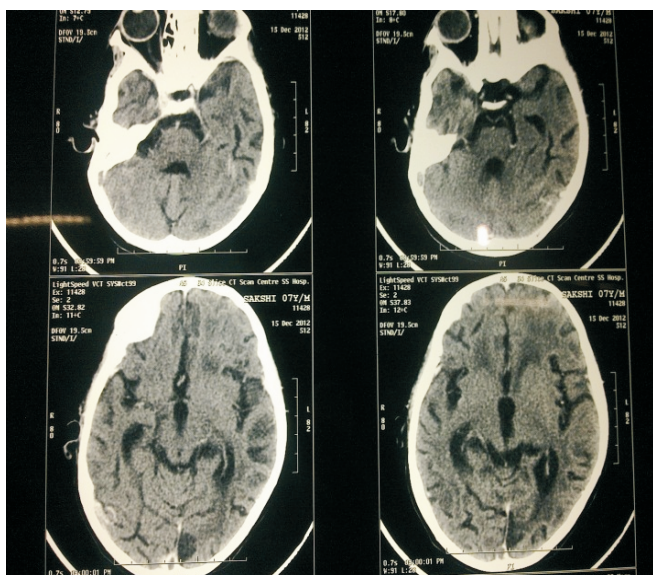
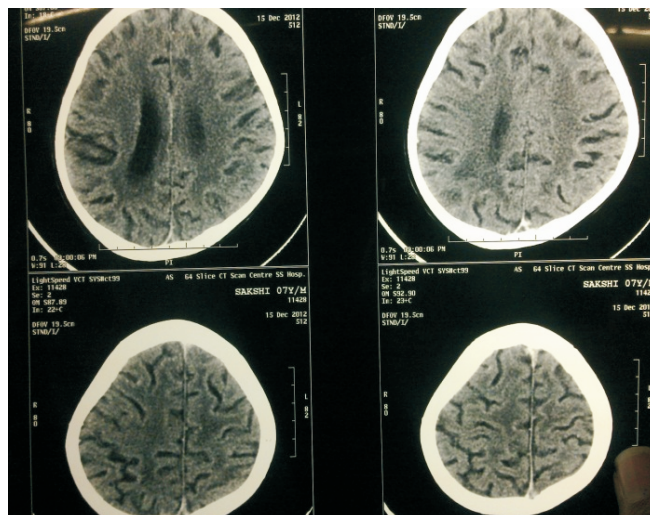


Figure 2: cranial tomography showing marked cerebral atrophy with prominent sulci and gyri at presentation to us.



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