Progressive Multifocal Leukoencephalopathy in a Case of Acute Lymphocytic Leukemia

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Progressive multifocal leukoencephalopathy (PML) is a rare neurologic condition which usually occurs in patients with leukemia, malignant lymphoma, carcinomatosis, acquired immunodeficiency syndrome (AIDS) or a variety of other chronic disease process or in those on immunosuppressive therapy(l). The diffuse parenchyma! infection is caused by a group B human polyoma virus serotype (JC virus)(2). In children, it is even more fatal than adults(3).

Case Report

A 7-year-old male child was diagnosed in this hospital 5 years back as a case of acute lymphatic leukemia and was treated accordingly. The child was readmitted two years and eleven months after the first admission with CNS and bone marrow relapse and treated with complete remission. On maintenance therapy with reinforcement he kept well for two years and then started having twitching of left side of face for 2-3 minutes on 2 consecutive days and intermittent difficulty of vision particularly while watching television for 3-4 days prior to the present admission. On examination, he was fully conscious, co-operative and oriented. He weighed 20 kg, his height was 118 cm and head circumference was 50 cm. Physical, systemic and ophthalmologic examination did not reveal any abnormality. The hemoglobin level was 12.4 g/dl, the total and differential counts were normal and peripheral smear showed normal morphology. CSF cytology and biochemistry were normal. The CSF culture was negative. EEG showed non-specific changes. One day after admission, the child had twitching of left side of face lasting for 15 minutes which was controlled with diazepam. However, he had 3 episodes of partial seizures again and carbamazepine was started. He had dilatation of left pupil with sluggish reaction to light, lower motor neurone type of weakness of left side of face and internal squint in left eye. Ophthalmoscopic examination revealed no abnormality. The CT scan of brain was normal. MRI *(Figs. 1&2)* showed multiple rounded lesions following the contours of gray white interface and showing outer scalloped margins in the right frontal, parietal and occipital regions. The lesions were hyperintense and there was no contrast enhancement. Similar lesion was seen in the right thalamus. The impression was progressive multifocal leukoencephalopathy (PML). ELISA test for AIDS was negative. The condition of the child deteriorated rapidly with development of generalized

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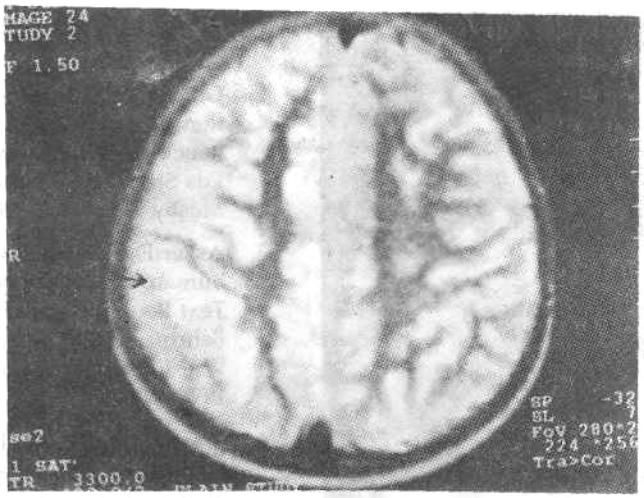

Fig. 1. MRI showing multiple rounded lesions following the contours of gray white interface and showing outer scalloped margins in the right frontal parietal and occipital regions.

seizures, left sided hemiparesis and deterioration of consciousness. Addition of clonazepam reduced the severity of convulsions. The child had respiratory tract infection and diarrhea. He expired three weeks after admission despite use of antibiotics and supportive therapy.

Discussion

PML is a diffuse parenchymal infection with very typical MR distribution^). The early lesions of PML are small, oval or round, begin in the subcortical white matter and spread to deeper white matter becoming large and confluent. There is less commonly gray matter involvement, and no mass effect. When the lateral margins of the lesion follow the gray-white interface, a scalloped appearance results. While PML classically occurs in a parieto-occipital distribution, it increasingly occurs in unusual location. "Atypical patterns" which have been observed in few patients with AIDS include thalamic and basal ganglia involvement(4).

In the present case, the diagnosis of PML is justified on the basis of clinical history and investigations particularly the typical MR distribution. Leukemic deposits in brain were ruled out as repeated CSF studies did not reveal leukemic cells, even after centrifugation(5). Leukemic cerebral masses and therapy induced sequalae in cases of ALL which include necrotizing leukoencephalopathy, mineralizing microangiopathy or diffuse cerebral atrophy are easily demonstrated by CT or MR images(6-8). The normal CT scan and typical MR distribution ruled out the possibility in the present case. Absence of fever, normal CSF cytology and biochemistry and negative culture finding exclude the possibility of other infective etiology. The CT scan or MRI did not reveal any exudate, abscess or other evidence of infection.

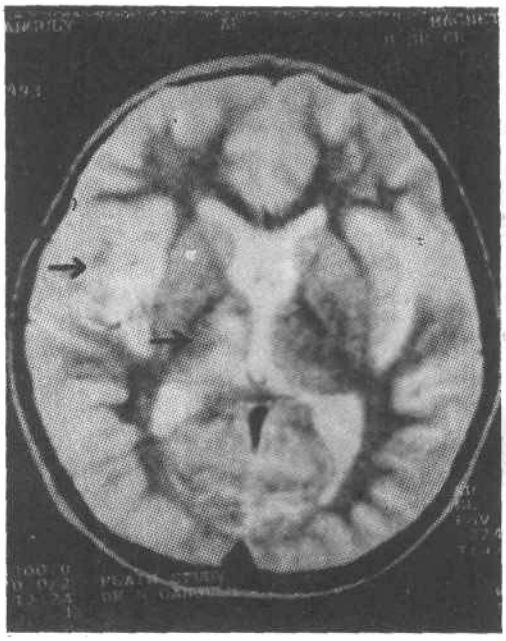

Fig. 2. *MRI showing hyperintense lesions at the gray white interface in the right parinetal region and the right thalamus.*

The virus in PML has not been distributed in tissues other than brain. The disease has not been transmitted to animals. There are isolated reports of clinical remission with cytarabine hydrochloride but no cure(l). Death usually occurs within 6 months of onset.

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