CASE REPORT

Primary CNS Lymphoma in Seropositive Child

S. U. Chakre [1], S. T. Bandichhode[1]*
[1]Department of Pediatrics, Dr. V. M. Government Medical College, Solapur - 413255 (Maharashtra), India

Abstract:

Primary Central Nervous System Lymphoma (PCNSL) is an extremely rare condition in childhood as compared to adults. It is seen in immunocompromised patients. We report the case of PCNSL in a child in Civil Hospital, Solapur which is a district anti-retroviral treatment (ART) centre. This 15 months old child presented with focal neurological deficit. The CT and MRI scan were suggestive of PCNSL. Stereostatic biopsy could not be done in this child. But CSF study showed 20 cells and all were lymphoblasts.

Keywords: Primary central nervous system lymphoma (PCNSL), immunocompromised patients, mass effect.

Introduction:

Primary Central Nervous System Lymphoma (PCNSL) is an extra nodal non-Hodgkin's lymphoma (NHL) arising from the brain parenchyma, eyes, meninges, or spinal cord in the absence of systemic disease [1]. It is a common complication of human immunodeficiency Virus-1 infection in adults [2]. But it is uncommon in pediatric patients with HIV-I infection. It forms approximately 1% to 3% of all central nervous system malignancies [1-4]. PCNSL has been diagnosed in HIV-positive children as young as 2 years [5]. Immunocompromised patients are at the particular risk for developing PCNSL such as individual affected with Human Immunodeficiency Virus (HIV), receiver of organ transplantation, or sufferer of congenital immunodeficiency syndromes. In this setting, PCNSL is due to Epstein-bar virus (EBV). Presentation of PCNSL depends upon the anatomic location of the lesions and presents more often as single brain lesion, either supratentorials (87%), and frontoparietal lobes (39%) [6].
The most common presentations included focal neurologic deficits (56% to 70%), mental status and behavioral changes (32% to 43%), symptoms and signs of increased intracranial pressure and seizures (11% to 14%) [6]. Multiple lesions are seen in 30% to 40% of the cases. Brain biopsy, lumbar puncture or vitrectomy can be used to establish tissue diagnosis [3]. Differential diagnosis on CT or MRI is toxoplasmosis and PCNSL. The masses are isodense or hypodense on CT and show contrast enhancement. On MRI, most lesions are hypodense on T_1 weighted, isodense or hyperdense on T_2 weighted images.

Case:

Our patient 15 months old female child 2nd product of third degree consangious marriage, wani by community, was brought by mother with fever, cough, cold and weakness in left side of body since 5 days. Patient was diagnosed as seropositive for HIV by DNA-PCR at 6 months of age. She was admitted one month back in our hospital for pulmonary tuberculosis. She was

started on anti-tubercular treatment and septran prophylaxis. A month later she developed weakness of left side of the body. Mother also complained of irritability. There was no history of convulsions and altered sensorium. Both parents were seropositive. They are not on anti retroviral therapy. The child was born normally at home and breast fed for a month. Later child was given mixed feeding. The child did not receive nevirapine and is immunized till date. The child had developmental delay and was below 3rd percentile for weight and height.

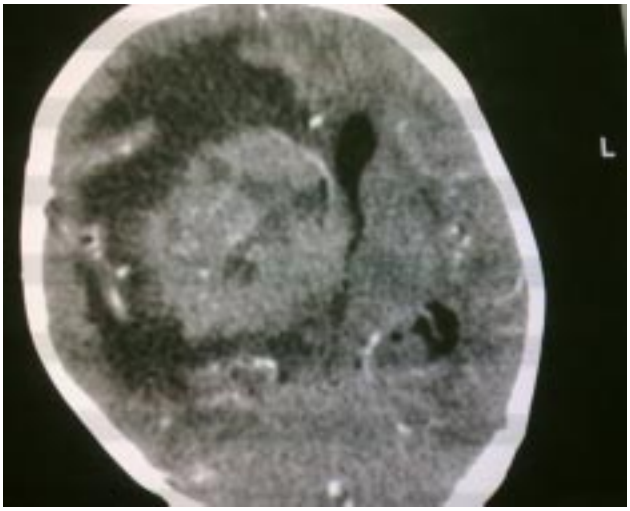
On examination, the child was febrile, HR ll8/min, RR 46/min; BP 80/60mm of Hg. Pallor was present. Anterior fontanellae was open. On CNS examination, the child was irritable and had left sided upper motor neuron type of facial palsy. Power was grade III on left side and normal on right side. Spasticity was present on left side. Deep tendon reflexes were brisk on left side and normal on right side. Plantar was extensor on left side and normal on right side. Sensory system was normal. Abdominal examination showed 3 cm; firm, non-tender liver and 3cm, firm Spleen. Cardiovascular system examination was normal. Crepitations were present on both lungs.

Investigations:

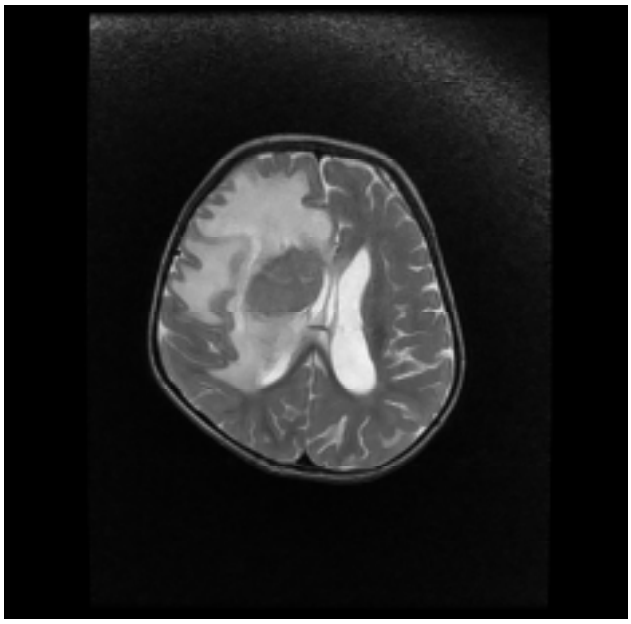
Complete Blood Count (CBC)-revealed Hemoglobin of 7.4g/dl, Total Lymphocyte Count of 4800/cmm, Differential Leucocyte Count (DLC) and Platelet counts are normal. CD4 count was 260/cumm (8%), and ESR was 32 mm at end of 1 hour. CSF examination showed 20 nucleated cells and all were lymphoblasts. CSF Biochemistry was normal. CSF culture was sterile. Chest X ray showed pneumonitis. Fundoscopy was normal. Toxoplasma serology was negative.

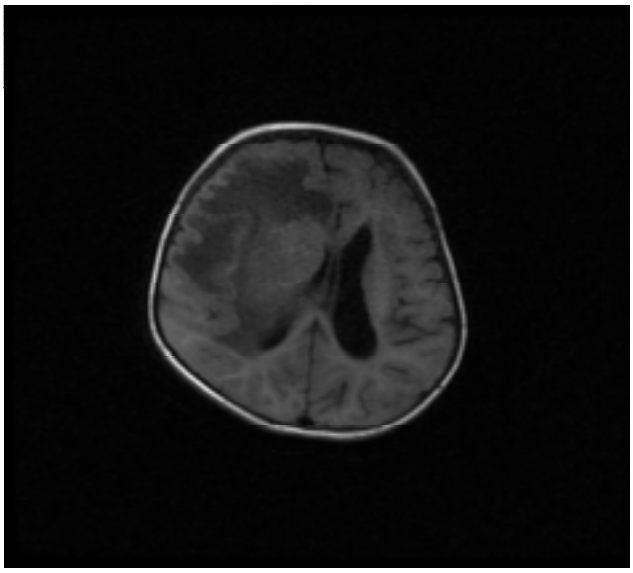
CT scan showed iso to hyperdense lesion of 4.8 ×4.2 ×3.5cm with contrast enhancement and adjacent edema involving right basal ganglia, thalamus, hypothalamus, caudate nucleus, adjacent right frontotemporal lobe and periaqiductal grey matter with ependymal extension and mass effect with dilatation of con-

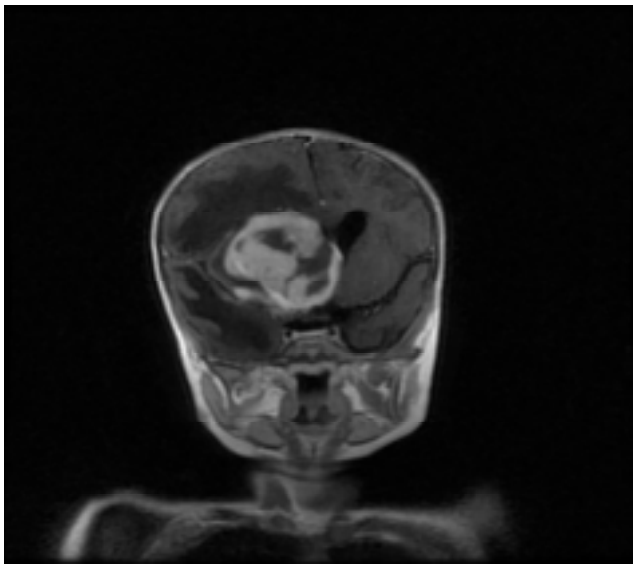
CT Scan



MRI Brain







tralateral ventricle and midline shift.

Intra axial neoplastic solid signal intensity lobulated mass was seen in the region of right basal ganglia and adjacent right frontal lobe white matter with perilesional edema and mass effect. On the basis of radio imaging, we thought of primary CNS lymphoma and toxoplasmosis as differential diagnosis. But considering seropositive status of the patient, negative serology for toxoplasma and toxoplasma being common in adult seropositive patients, primary CNS lymphoma was the most likely possibility. Because of unavailability of the speciality services at our centre, we referred this child to higher centre in Mumbai, but this patient expired during treatment.

Discussion:

75 % of the patients with PCNSL have advanced HIV disease. Although PCNSL has been seen in adults and older children, its presence in an infant is rare [1, 2, 4, 7-9]. Our patient presented with focal neurological deficit. Thinking of it as an opportunistic CNS infection, we have done fundoscopy and lumber puncture. CSF has shown 20 cells and all were lymphoblasts. Then CT scan has shown single mass lesion with contrast enhancement. CT thorax has not shown lymphadenopathy. Biopsy could not be done in our patient. So, we have MRI brain which showed supratentorial periventricular mass. Other tumorus occurring in childhood or adolescence such as primitive neuroectodermal tumours may have similar appearance on CT/ MRI. In HIV infected patients, PCNSL is the most likely diagnosis. On the other hand, Toxoplasmosis is the most common cause of space occupying lesion of the brain in adults with HIV infection. CNS toxoplasmosis is exceedingly rare in young infants, but may occur in HIV-infected adolescents we can diagnose PCNSL by radio imaging, but needs to be confirmed before starting treatment. CNS irradiation and oral prednisone are treatments for lymphoma and may prolong survival. Success of treatment for lymphoma is disappointing. Despite intervention, prognosis is poor; however, Highly Active

Anti-Retroviral Therapy (HAART) seems to improve the prognosis in several cases.

References:

1. Shiozawa Y, Kiyokawa N, Fujimura J, Suzuki K, Yarita Y, Fujimoto J, Saito M, Yamashiro Y. Primary malignant lymphoma of the central nervous system in an immunocompetent child: a case report. *J Pediatr Hematol Oncol* 2005; 27(10):561-4.

2. Cecchi PC, Billio A, Colombetti V, Rizzo P, Ricci UM, Schwarz A. Primary high-grade B-cell lymphoma of the choroid plexus. *Clin Neurol Neurosurg* 2008; 110(1):75-9.

3. Nimish A, Moile MD, Lauren E, Abrey MD. Primary central nervous system lymphoma. Neurologic Clinics 2007; 25:1193-1207.

4. Scrigni A, Nastri M, Rodiguez de Schiavi s, Czornyi L, Felice M, Manrese B. Leptomeningeal lymphoma in a child with acquired immune deficiency syndrome. Neuropediatrics 2006; 37(3):121-5.

5. Fitzsimmons A, Upchurch K, Batchelor T. Clinical Features and Diagnosis of Primary central nervous system lymphoma. *Hematol Oncol Clin North Am* 2005; 19(4):689-703, vii.

6. Iwamoto FM, DeAngelis LM. An update on primary central nervous system lymphoma. *Hematol Oncol Clin North Am* 2006; 20(6):1267-85.

7. Karikari IO, Thomas KK, Lagoo A, Cummings TJ, George TM. Primary cerebral ALK-1-positive anaplastic large cell lymphoma in a child. Case report and literature review. *Pediatr Neurosurg* 2007; 43(6):516-21.

8. Doolittle ND, Abrey LE, Shenkier TN, Tali S, Bromberg JE, Neuwelt EA, Soussain C, Jahnke K, Johnston P, Illerhaus G, Schiff D, Batchelor T, Montoto S, Kraemer DF, Zucca E. Brain parenchyma involvement as isolated central nervous system relapse of systemic non-Hodgkin lymphoma: an International Primary CNS Lymphoma Collaborative Group report. Blood 2008; 111(3):1085-93.

9. Medina- Sanson A, Chico-Ponce de Leon F, Cabrera- Munoz Mde L, Gallegos-Castorena S, Caltenco-Serrano R, Barragan- Perez E. Primary central nervous system non-Hodgkin lymphoma in childhood presenting as bilateral optic neuritis. *Childs Nerv Syst* 2006; 22(10):1364-8.

***Author for Correspondence:** *Dr. S. U. Chakre, 22, Narayanprasad, Krushinagar Civil lines, Solapur-413003 (Maharashtra). Mobile: 9049883035*
Email: drgschakre@yahoo.co.in.