HIV INFECTION PRESENTING AS STROKE AND ASYMPTOMATIC DANDY-WALKER MALFORMATION IN AN ADULT

Amit Agrawal¹, A. N. Hegde², Lathika Shetty³, Boby Varkey³, J.P. Shetty⁴

K.S.Hegde Medical Academy, Departments of Neurosurgery¹, Internal Medicine² Radiology³ and Pathology⁴, Mangalore, India

We report a unique and rare case with definite human immunodeficiency virus infection whose initial presentation was stroke and associated finding of asymptomatic Dandy-Walker malformation in an adult.

Key words: HIV, cerebral infarction, stroke, Dandy-Walker malformation

Eur J Gen Med 2005; 2(3):135-137

INTRODUCTION

Human immunodeficiency virus (HIV) infection is a devastating viral infection and neurological dysfunction as the first manifestation of HIV has been found in 10 to 20% of symptomatic patients (1,2). The incidence of clinically evident cerebral infarction in AIDS (Acquired immunodeficiency syndrome) patients varies between 0.5% and 7% (3-5). However stroke as the first presentation of the illness is rare (6). We report a unique case of an adult that presented with stroke as the first sign of AIDS and a rare association of asymptomatic Dandy-Walker malformation.

CASE

A 40 year female presented with fever of 4 days and sudden onset weakness of right side of body since 2 days. There was no previous history of smoking, systemic arterial hypertension, diabetes mellitus or any significant illness. Her husband was a lorry driver. On examination pallor was present and other general and systemic examination was unremarkable. Neurologically she was conscious, alert and oriented. Motor examination revealed mild weakness of the right side of her face, weakness of grade 3/5 involving right upper and lower limb with hyperreflexia and positive Babinski's

sign bilaterally. There were no meningeal signs. Complete blood count revealed Hb-8.6 gm/dl, total white cell count -3,100/mm³ with polymorphs 60%, lymphocytes 38% and eosinophils 2%. Erythrocyte sedimentation rate was 145 mm/Hr. Peripheral blood smear demonstrated microcytic hypochromic red blood cells with an adequate amount of platelets. Blood chemistry and liver function tests were within normal limits. Urinalysis was normal. Antinuclear antibodies, rheumatoid factor, and lupus cell preparation were all normal. Coagulation and protein C were within normal limits. A lumbar puncture was performed, which was unremarkable. Routine Gram stain of CSF and bacteria culture were negative. CSF VDRL was negative. Her serologic tests revealed positive HIV antibody. CT scan of the brain revealed an infarct involving posterior limb of internal capsule (Figure 1). It also showed hypoplasia of cerebellar vermis and large posterior fossa cyst communicating with the enlarged fourth ventricle suggestive of Dandy-Walker malformation and dilated third and lateral ventricles (Figure 2). However sulci and gyri were well seen. She was managed conservatively and started on antiretroviral therapy. She is doing well and hemiplegia improved minimally.

E. mail: dramit in@yahoo.com

136 Agrawal et al.

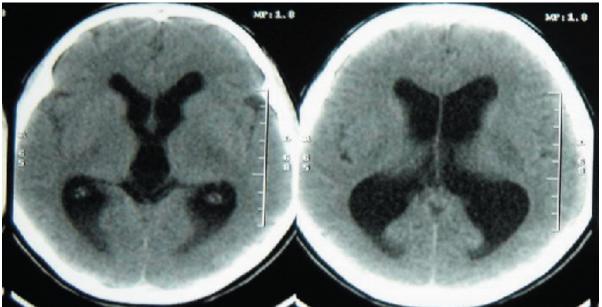


Figure-1. CT scan without contrast showing hypodensity in the posterior limb of left internal capsule compatible with an acute infarction with dilated third and lateral ventricles. Bilateral Sylvian cisterns are well seen.

DISCUSSION

The cause of cerebral infarction or transient neurological deficits in AIDS patients can be found in approximately 50% of cases (7). The most common causes of cerebral infarction in AIDS are nonbacterial thrombotic endocarditis and opportunistic central nervous system infections i.e. toxoplasmosis, cryptococcal

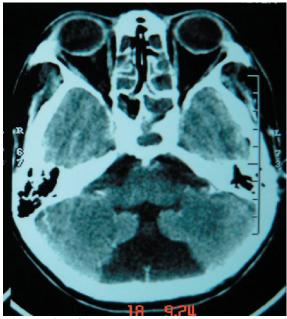


Figure-2. CT scan showing agenesis of cerebellar vermis, large posterior fossa cyst communicating with the fourth ventricle suggestive of Dandy-Walker malformation

meningitis and tuberculosis (2,3,8). However in a good majority of the cases no identifiable cause could be found and the pathogenesis remains unclear (9). It is described that in these patients potential vascular mechanisms cerebral infarction and transient neurological deficits include a direct toxic effect of viral antigen or vasculopathy caused by HIV (3,10). In our patient no clue was evident indicating impaired immune function up to the onset of hemiparesis. With the available tests, no evidence was present of an inflammation or infection that might be responsible for these ischemic events. No underlying metabolic or cardiovascular disorders were found. Because of the nonreactive VDRL in her CSF, cerebral infarction caused by treponema was unlikely. Therefore we conclude that stroke in this patient was most likely caused by HIV. However, it is not possible to postulate a specific pathogenesis for this ischemic infarction. We presume that ischemic infarction was secondary to vasculopathy caused by HIV. Associated Dandy-Walker malformation in this patient was an incidental finding and she was asymptomatic for that since childhood. Dandy-Walker syndrome is a rare congenital malformation of the hindbrain involving the cerebellum and the fourth ventricle that commonly occurs before the sixth or seventh week of development. It is usually diagnosed at birth or in early childhood. Adult presentation of the Dandy-Walker syndrome is extremely rare (11,12). We could find only one case report where patient never developed evidence of cerebral involvement till adulthood and Dandy-Walker syndrome was an incidental finding (13). This case is unique and rare where the patient presented with ischemic infarct as a first manifestation of HIV infection and also had asymptomatic Dandy-Walker malformation. From the management point of view she is planned to continue on anti-retroviral therapy and close follow-up for Dandy-Walker malformation.

REFERENCES

- Mcguire D, So YT. Neurological dysfunction: overview. In: Cohen PT, Sande MA, Volberding PA, eds. The AIDS Knowledge Base. 2nd ed. Boston, Mass: Little Brown & Co;1994:5.6-1,5.6-2
- 2. Pinto AN. AIDS and Cerebralvascular Disease. Stroke 1996;27(3);538-43
- 3. Engstrom JW, Lowenstein DH, Bredesen DE. Cerebral infarctions and transient deficits associated with acquired immunodeficiency syndrome. Am J Med 1989;86:528-32
- 4. Gillams AR, Allen E, Hrieb K, et al. Cerebral Infarction in Patients with AIDS. AJNR 1997;18:1581-5
- 5. Levy RM, Bredesen DE, Rosenblum ML. Neurological manifestations of acquired immunodeficiency syndrome (AIDS) at UCSF and review of the literature. J Neurosurg 1985;62:475-95
- Visudtibhan A, Visudhiphan P, Chiemchanya S. Stroke and seizures as the presenting signs of HIV infection. Pediatr Neurol 1999;20:53-6

- 7. Saul RF, Gallagher JG, Mateer JE. Sudden hemiparesis as the presenting sign in cryptococcal meningoencephalitis. Stroke 1986;17:753-4
- 8. Picard O, Brunereau L, Pelosse B et al. Cerebral infarction with vasculitis due to varicella zoster virus in patients infected with the human immunodeficiency virus. Biomed Pharmacother 1997;51:449-54
- 9. Brew BJ, Miller J. Human immunodeficiency virus type I related transient neurological deficits. Am J Med 1996;101:257-61
- 10. Leite AGB, Vidal JE, Filho FB, Nogueira RS, de Oliveira ACP. Cerebral infarction related to cryptococcal meningitis in an HIV-infected patient: case report and literature review. Braz J Infect Dis 2004;8(2)
- 11. Freeman SR, Jones PH. Old age presentation of the Dandy-Walker syndrome associated with unilateral sudden sensorineural deafness and vertigo. J Laryngol Otol 2002;116(2): 127-31
- Reddy P, Vannemreddy PS, Grier L, Nanda A. Dandy-Walker syndrome: presentation of the congenital malformation in an older patient. La State Med Soc 2000;152(1): 31-4
- 13. Yamamoto Y, Waga S. Asymptomatic Dandy-Walker variant in an adult. Surg Neurol 1984;22(3):241-2