WEBER'S SYNDROME DUE TO CEREBRAL TOXOPLASMOSIS

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Introduction

Weber's syndrome is the occurrence of third nerve palsy with contralateral hemiplegia, usually due to a cerebrovascular accident involving the proximal precommunal part of the posterior cerebral artery [1]. Other causes of the syndrome include opportunistic infections, brain tumours, vascular aneurysms & demyelination. Though, *Toxoplasma gondii* is one of the commonest pathogens causing opportunistic cerebral infection in patients with acquired immunodeficiency syndrome (AIDS), brainstem dysfunction due to this infection is rare [2-4]. Besides toxoplasmosis, several other malignancies & infections such as lymphoma, progressive multifocal leucoencephalopathy (PML) and cytomegalovirus (CMV) infection can also result in brainstem involvement in these patients [5-7].

We report one such case of Weber's syndrome due to cerebral toxoplasmosis & an uncommon stroke-like presentation in this patient.

Case Report

A 45-years-old, right handed lady, presented with sudden onset right sided hemiparesis, diplopia on looking to the right, drooping of left upper eyelid and facial asymmetry. She did not give any history of transient ischemic attacks, hypertension or features of raised intracranial tension (ICT) at presentation. There was no history of extramarital sexual exposure, intravenous drug abuse or any sexually transmitted disease. On examination she was normotensive & there were no neurocutaneous markers. Neurological assessment revealed a fully conscious & alert patient with no higher mental function disturbance. Her speech was normal at presentation. There was clinical evidence of left oculomotor nerve palsy in the form of ptosis of the left eye, medial rectus palsy and pupillary dilatation (Figs 1 & 2). She also had right sided hemiparesis & right facial nerve palsy of an upper motor neuron type. Examination of other systems was within normal limits. On investigations, routine hemogram & biochemical parameters were normal. A CT scan (Head) showed an area of low attenuation in the left brainstem region (Fig 3) with no evidence of raised ICT. In view of the acute stroke like presentation involving the posterior circulation, prophylactic management with anti cerebral edema measures (oral glycerol, 30 ml 8th hourly) & anti-

platelet drugs (Tab aspirin, 325 mg, once daily) was started. Other causes of hypo-attenuated lesion on CT Scan including cerebral edema or cerebritis were not considered at this stage due to the clinical picture. Despite therapy, she continued to deteriorate with a worsening of hemiparesis. Two weeks later, she developed obtunded sensorium & persistent projectile vomiting due to raised ICT. An MRI scan was done at this stage which revealed two ring enhancing lesions in the left cerebral peduncle & internal capsule (Fig 4 & 5) along with features of raised ICT in the form of midline shift & compression of left lateral ventricle. At this stage, her serum was tested for IgM anti-toxoplasma antibodies and was reported as positive. ELISA for HIV-1 was positive on two occa-

Fig. 1 : Oculomotor palsy leading to ptosis of the left eye in a case of Weber's syndrome due to cerebral toxoplasmosis

Fig. 2 : Same patient showing lateral deviation of the left eye due to an un-opposed action of the lateral rectus

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Fig. 3: CT scan head of the same patient showing a hypo-attenuated area in the region of the left basal ganglia and brain stem

Fig. 4: MRI scan of the patient showing ring enhancing lesions in the region of left cerebral peduncle

sions. Cerebrospinal fluid (CSF) analysis revealed mildly elevated proteins (70mg/dl) with mild lymphocytic pleocytosis (15 lymphocytes/cmm). CSF serology for IgM anti-toxoplasma antibodies was positive while it was negative for HIV & VDRL. The patient's husband also tested positive for HIV antibodies by ELISA. She was managed with sulphadoxime-pyrimethamine combination (2g/75 mg per day in divided doses) along with aggressive anti cerebral edema measures (Inj mannitol, 20%, 200ml, IV, 8th hourly & oral glycerol, 30 ml 8th hourly). She had a satisfactory response to therapy with a regression of features of raised ICT & a halt in the progression of the neurological deficit. She was discharged after two weeks of therapy with advice to continue life-long prophylactic sulphadoxime-pyrimethamine & health education regarding life-style modification.

Discussion

Although subclinical brainstem involvement by hu-

Fig. 5: MRI scan of the patient showing ring enhancing lesions in the region of midbrain

man immunodeficiency virus (HIV) has been documented by brainstem auditory evoked potentials [8], symptomatic brainstem involvement in such cases usually indicates an opportunistic infection or a neoplasm. Lymphoma is the commonest tumour involving the central nervous system (CNS) in AIDS patients. However, brainstem is an uncommon site for these tumours as suggested by a paucity of such reports in literature [5,6]. Infective causes of brainstem involvement in HIV disease include progressive multifocal leukoencephalopathy (PML) [7,9], herpes zoster [5,10], cytomegalovirus infection [11] & cryptococcal meningitis [6,13].

Cerebral toxoplasmosis, is the commonest CNS opportunistic infection in AIDS patients [2,14]. Despite this, few cases of brainstem involvement due to toxoplasmosis are reported in literature. Isolated case reports include one patient with third nerve palsy & contralateral ataxia (Claude's syndrome) [4], two with ocular motility defects [12] and 2 with an oculomotor nerve palsy & rubral tremor [15,17]. In a prospective analysis of ophthalmological manifestation of AIDS [13], toxoplasmosis was the etiological agent in only one out of a total of 26 patients with cranial nerve palsy. Analysing differently, Navia et al found only one patient with brainstem involvement in their series of 27 cases with CNS toxoplasmosis [3]. In one of the larger retrospective analyses, brainstem toxoplasmosis was reported in 8 out of a total of 366 patients with AIDS & CNS toxoplasmosis [16]. The basis of diagnosis in the above study was either an improvement in symptomatology after institution of appropriate therapy or histological confirmation of the diagnosis. Six out of these 8 patients had oculomotor palsy and con-

tralateral hemiplegia (Weber's syndrome) with or without additional findings such as dysarthria (1 patient), seizures (1) rubral tremor (1) & bilateral corticospinal tract signs [1]. CT scan revealed enhancing lesions in all these cases & hence MRI was not done in any of them. An acute stroke like presentation -as seen in our patient- is extremely rare in cerebral toxoplasmosis & has been described by Navia et al [3]. The pathological cause of stroke & the resultant neurological deficit in these patients could be vasculitis or a direct invasion of the vascular supply of the involved territory.

The present case therefore demonstrates three important clinico-radiological considerations. First, it documents the occurrence of brainstem toxoplasmosis, which is a rare site of cerebral toxoplasmosis. Secondly, a stroke like presentation, as documented in this case, is even rarer & can result in some delay in the diagnosis & initiation of appropriate therapy. Finally, the superiority of MRI over CT Scan in diagnosing parenchymal lesions of the brain-particularly in the posterior fossa is well highlighted by this case.

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