CASE REPORT

Thalamic Pain Syndrome Due to Cytomegalovirus Vasculitis in an HIV- Positive Child

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Dejerine-Roussy syndrome, also known as the 'thalamic pain syndrome' is a condition in which the body becomes hypersensitive to pain as a result of damage to the thalamus, a part of the brain that affects sensation. Association of this syndrome with HIV is rare with few case reports described in adults. We report a 10-year-old male child who was HIV positive and had developed this syndrome due to cytomegalovirus vasculitis.

Key words: *Cytomegalovirus vasculitis, Dejerine Roussy syndrome, Human immunodeficiency virus, Thalamic pain syndrome.*

Dejerine-Roussy syndrome, also known as thalamic pain syndrome is a condition associated with inadequate blood supply from the posterior cerebral artery. It is characterised by prominent and persistent sensory loss that affects both superficial (touch, pinprick and temperature) and deep (vibration and position sense) sensations, slight or transient hemiparesis and intractable aching or burning pain on the affected side of the body with a thalamic lesion on the opposite side(1-3). Thalamic pain syndrome due to cytomegalovirus (CMV) vasculitis in an HIV-positive child has not been described previously. We report a 10-year-old male child who presented with this condition.

CASE REPORT

A 10-year-old boy was admitted with complaints of pain and gradual onset weakness in the left upper and left lower limb, since 10 days. The pain was constant and burning in character. Allodynia (a painful response to a usually non-painful stimulus) was also present. There was a history of excessive sleepiness for 8 days and fever for 2 days. There was no history of contact with tuberculosis. Both his parents were HIV-infected and he was diagnosed as HIV-infected 6 months back by enzyme-linked immunosorbent assay (ELISA) method, when his CD4 count was 102 cells/cumm. Cotrimoxazole prophylaxis and anti-retroviral therapy (ART) with stavudine (2 mg/kg/day), lamivudine (8 mg/kg/day) and nevirapine (300mg/m^2/day) was started. There was no history of blood transfusion or admission in the past. The child's mode of acquisition of HIV was most probably through perinatal transmission.

On admission, the child was afebrile with a pulse rate of 100/min, respiratory rate 20/min and blood pressure 100/70 mm of Hg. His weight was 22.5 kg and height was 133cm (<5th percentile for age). The child was drowsy and fundus examination showed evidence of retinitis. Visual acuity was normal. Cranial nerve examination was normal. The tone in the left upper and left lower limbs was decreased with power of 2/5. Tone and power were normal on the right side. Deep tendon reflexes in all four limbs and superficial reflexes were normally elicited. Sensory system examination revealed loss of both superficial (response to touch, pinprick and temperature) and deep (vibration and position sense) sensations in the left upper and lower limb. Sensation over the right upper and lower limbs was

normal. There were no meningeal or cerebellar signs. Other systemic examination was normal.

Investigations showed hemoglobin of 10.4 g/dL, white blood cells of 7600/cumm (neutrophils 63%, lymphocyte 33%, and monocytes 4%) and platelet count 334 × 10^3 /cumm. Random blood sugar, liver and renal function tests, serum electrolytes and serum calcium were normal. The Mantoux test was negative and chest *X*-ray was normal. Magnetic resonance imaging (MRI) of the brain revealed an ill-defined area of altered signal intensity in the right thalamic and sub-thalamic region suggestive of ischemia. The cerebrospinal fluid examination was normal. Polymerase chain reaction of CSF for cytomegalovirus was positive. Serology was positive for both CMV IgM and IgG. His titres for varicella IgG and IgM and Toxoplasma IgM and IgG were negative. A diagnosis of thalamic pain syndrome secondary to CMV vasculitis was made based on the history, examination, presence of CMV retinitis, positive serology test for CMV and findings on neuroimaging. He was started on intravenous ganciclovir (5 mg/kg/dose, twice daily) following which there was a gradual improvement in power on the left side. Gabapentin and amitriptyline was started for his pain. Stavudine was replaced with zidovudine (240 mg/m^2/dose) in view of CNS involvement, and lamivudine with nevirapine was continued. At completion of 21 days of ganciclovir, power in the left upper and lower limb had returned to 5/5 and sensory system examination was normal. He was discharged on oral ganciclovir for secondary prophylaxis (30 mg/kg/day) and on antiretroviral therapy. He is well on follow up after 2 months with no neurological deficit. His CD4 count on follow up was 1070 cells/cumm and ophthalmological examination revealed resolution of the lesions of CMV retinitis.

DISCUSSION

Dejerine - Roussy syndrome was originally described by Dejerine and Roussy in 1906, and attributed to infarction in the thalamus(4). The thalamus has been described as the sensory relay station of the brain. Pain or discomfort may be felt on the affected side after being mildly touched or even in the absence of a stimulus. The pain may aggravate by exposure to heat or cold and by emotional distress. Various sensory abnormalities are commonly found in these patients, including disturbances of temperature discrimination(6).

This syndrome has been described in adults secondary to unruptured cerebral aneurysm, stroke and cerebral abscess(1-3). Gonzales has described it in an HIV-positive patient due to toxoplasmosis(2). There are a few case reports in children, which describe this syndrome secondary to measles infection and tuberculous meningitis(7-8).

Almost every pattern and type of vasculitis of small, medium and large vessels has been encountered in the HIV setting. It can range from vasculitis resulting from specific infective agents to a non-specific vasculitis. The common infective causes include cytomegalovirus and tuberculosis(9). The major mechanisms by which infection is thought to induce vasculitis, are direct microbial invasion with resultant damage of the vessel wall, and immune-mediated injury (both humoral and cellular). The organs usually involved are skin, peripheral nerve, skeletal muscle and the central nervous system(9).

In our patient, the thalamic lesion was probably due to CMV vasculitis since PCR for CMV was positive and symptoms improved with ganciclovir therapy. Vasculitis has been proposed as a cause of thalamic syndrome secondary to measles infection(7). Various hypotheses for the mechanisms of pain in thalamic syndrome have been proposed. It could result from an "irritable focus" at the central site of injury or from a lack of inhibition in the pain-signaling system, i.e. the spinothalamic pathways(6).

Another theory suggests that the pain is due to disruption of thermosensory integration, the loss of cold inhibition resulting in the experience of burning pain. The lesion in the spinothalamic projections, removes the suppressing activity exerted by the reticular thalamic nucleus, thereby releasing abnormal activity in other thalamic nuclei, which in turn leads to pain hypersensitivity(6).

Pain in thalamic pain syndrome is treated with antidepressants, especially those which are adrenergically-active (*e.g.* amitriptyline). These should be started as soon as the diagnosis is

suspected. Response to therapy usually occurs within four weeks. In our patient the response was seen after two days of starting the therapy. If the response is poor, carbamazepine or mexiletine may be useful(6).

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