Correspondence

Tropical Spastic Paraparesis from Northern India

Sir,

In 1964, a progressive myelopathy involving the thoracic region was termed Tropical Spastic Paraparesis (TSP). Subsequently a similar myelopathy associated with HTLV-1 antibodies in both serum and CSF patients were found and named HAM (HTLV-1 associated myelopathy). The epidemiological, clinical and pathological features of cases from India are similar to the description form rest of World, though attempts to link TSP and HTLV-1 have largely been unsuccessful. Most of the cases of TSP reported from India are from southern parts of the country. Although sporadic case reports have been made from various other parts, we report an interesting case having features consistent with TSP from northern India.

AK, 33-year male hailing from Motihari, Bihar presented to Medical OPD with a 13-year long duration illness. He had insidious onset, gradually progressive weakness of lower limbs beginning almost symmetrically. Lately from last 4 years he is bedridden with frequent flexor and extensor spasms. He gave history of bladder dysfunction (urinary frequency, hesitancy, urgency) beginning early in course of illness. For last 9 years the patient is having lower limb parasthesia (painless). There was no history suggestive of upper limb, cranial nerve or higher mental function abnormalities. The patient denied any history of Khesari dal intake, remission or acute exacerbation in symptoms, blood transfusion, high risk behaviour for STDs and relative or neighbour suffering from such illness. Examination revealed markedly increased tone with grade 0/5 power in both lower limbs with exaggerated deep tendon reflexes, sustained ankle clonus, bilateral extensor plantars and mildly impaired vibration sense in lower limbs. Higher mental function, cranial nerves, optic fundi, upper limbs and cerebellar functions were normal. Routine hemogram, blood biochemistry and X-ray of thoracolumbar and cervical spine were unremarkable. ELISA for HIV-1 and HIV-2 was negative. CSF analysis revealed 15 cells/mm³ all lymphocytes, normal glucose and protein with negative VDRL and absence of oligoclonal bands. MRI revealed atrophy of lower part of spinal cord. (Fig. 1).

The temporal profile of illness in the present case including typical clinical presentation, CSF and MRI findings are highly suggestive of TSP. However, HTLV-1 and HTLV-2 serology could not be carried out due to non-availability of the test, very high cost and low diagnostic yield specially in India. In a study from South India, out of 25 subjects only one case tested positive for HTLV-1 antibodies. The other differential diagnosis to be considered in such clinical syndromes include lathyrism, hereditary spastic paraplegia, spinal from of primary progressive multiple sclerosis, HIV related myelopathy and primary lateral sclerosis. Lathyrism can be differentiated from TSP by specific geographic distribution, khesari dal intake, other family members suffering from similar illness and frequent upper limb involvement. Hereditary spastic paraplegia is unlikely because it usually has a positive family history, young age of onset, marked spasticity with minimal pyramidal weakness, marked lumbar lordosis and acellular CSF. The spinal form of primary progressive multiple sclerosis may behave exactly like TSP. But absence of oligoclonal bands in CSF and characteristics MRI do not favour the diagnosis of MS. Bladder dysfunction, sensory symptoms, absence of pseudobulbar palsy and abnormal MRI are against the diagnosis of primary lateral sclerosis. HIV related myelopathy is unlikely in view of prolonged duration of illness, absence of other manifestations and negative serology.

Although the diagnosis of TSP is significantly supported by HTLV serological studies, the clinical recognition of this entity along with consideration of other differential diagnosis (as discussed above) is of
utmost importance in a country like ours where cost of such serological investigations may be prohibitive. This is backed up by the fact that most of Indian studies have failed to demonstrate this association and leading to possibility of seronegative spinal spastic paraparesis.  

In our case the clinical and temporal profile of the illness along with MRI findings were highly suggestive of TSP. Various treatment modalities like steroid, interferon-α, vitamin C, zidovudine, azathioprine, cyclophosphamide, IVlg have been tried but results are largely disappointing.  

The absence of serological evidence of HTLV infection may be due to infection by other viruses, nutritional factors or toxin exposure but the explanation remains elusive and we suggest to embark upon the clinical recognition of this rare entity.

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REFERENCES

Atypical Presentations of Sheehan’s Syndrome

Sir,

Sheehan’s syndrome, first described in 1937, is an adrenal pituitary insufficiency from hypovolemia secondary to excessive blood loss during or after delivery. It may present in post-partum period or several years after delivery. Patients of Sheehan’s syndrome present in emergency due to situations like coma, hypothyroidism, hypoglycemia or hyponatremia following a stressful event. Patients also presents with anemia, dry and light colour skin. Ozkan and Colak reviewed 20 cases of Sheehan’s Syndrome; 3(15%) presented with hypoglycemia, 3(15%) with hypothyroidism, 1(5%) with hyponatremia, 6 had empty sella and 9 had partial empty sella.

We report two cases of Sheehan’s syndrome with uncommon presentations - paralytic ileus and acute psychosis. The first patient was a 32 years female; presented with loose motions, vomiting and giddiness. On examination, she had fever, pallor, dry coarse skin, hoarse voice and madarosis, BP of 80/60 mm Hg; peristaltic sounds were absent. Serum Na+ 127 mEq/L, K+ 3.9 mEq/L, BSL® was 61mg%. X-ray abdomen erect showed multiple air fluid levels. Provisional diagnosis of acute gastroenteritis with paralytic ileus was made and treated accordingly. Peristalsis returned and air fluid levels on X-ray disappeared after 2 days. Repeat serum Na+ and K+ were 137mEq/L and 4.5mEq/L. Serum T₄, T₃, TSH levels were done, all of which were low. Obstetric history was reviewed; she had history of post partum hemorrhage 13 years ago followed by lactation failure, amenorrhea and loss of secondary sexual characteristics. She also gave history of increased lassitude, giddiness, vague indigestion and fatigue. Serum cortisol levels were low. Due to financial constraints, other hormonal levels could not be done. A diagnosis of Sheehan’s syndrome was made and she was put on oral steroids, followed by L-thyroxine and estrogen supplements.

The second patient was a 25 years female presenting with acute psychosis. She had multiple episodes of altered sensorium with frequent loss of consciousness since past one year. There was history of postpartum hemorrhage five years ago followed by amenorrhea. Her investigations one year ago showed subnormal levels of serum FSH, LH, HPRL, cortisol, T₃, T₄ and TSH. She was receiving oral L-thyroxine for hypothyroidism since then. One week ago she had been put on 60mg of oral prednisolone daily. On examination she was intermittently rowdy; had retrograde amnesia. A diagnosis of Sheehan’s Syndrome with acute psychosis was made. She was treated with antipsychotic drugs; dose of prednisolone was tapered slowly over a week and maintained at 7.5 mg/day. Thyroxine and estrogen supplements were given. In both the patients, USG abdomen-pelvis showed small atrophic uterus with shrunk ovaries and CT scan brain showed partially empty sella. The psychosis recovered within a week and after 10 days of presentation, she was off antipsychotic medications.

In the first patient, paralytic ileus despite normal serum K+ levels was due to hypothyroidism with acute gastroenteritis. Pseudo-obstruction of the intestine can occur in conditions like scleroderma, myxedema, diabetic autonomic neuropathy and amyloidosis. However, after extensive review of literature we have not come across any report of paralytic ileus as the presenting feature in Sheehan’s syndrome. We would like to highlight how we investigated the patient with high index of suspicion and arrived at the diagnosis and also emphasize the importance of a detailed past and obstetric history which sometimes takes a back seat in acute emergencies. In the second case, while the patient was on treatment outside, low TSH and cortisol levels were overlooked. Ideally, steroid replacement should precede thyroxine replacement. Also the recommended dose of steroids in a hemodynamically stable patient is 5 mg A.M and 2.5 mg P.M. Behavioral disorders can occur with high dose of steroids which probably happened in our case.