Erb's paraplegia with primary optic atrophy: Unusual presentation of neurosyphilis: Case report and review of literature

Chandramohan Sharma, Kunal Nath, Banshi LaL Kumawat, Dinesh Khandelwal, Deepak Jain

Department of Neurology, Sawai Man Singh Medical College, Jaipur, Rajasthan, India

Abstract

Symptomatic neurosyphilis (NS) can have varied syndromic presentations: Meningitis, meningovascular and parenchymatous involvement. Non-tabetic syphilis affecting the spinal cord is rare and only sporadic case reports have been published. The variant of meningomyelitis known as Erb's paraplegia refers to patients of spinal syphilis with very slow progression over many years and predominantly motor signs. Primary optic atrophy occurs twice as frequently in tabes dorsalis as in other types of NS. We describe here a case of a 32-year-old truck driver who presented with Erb's paraplegia with primary optic atrophy. This clinical overlap in NS is extremely rare and to our knowledge is the first case report of its type.

Key Words:

Erb's paraplegia, neurosyphilis, optic atrophy

For correspondence:
Dr. Kunal Nath, Senior Resident, Department of Neurology,
Sawai Man Singh Medical College, Jaipur- 302 004, Rajasthan, India.
E-mail: lanukhtan82@yahoo.in

Ann Indian Acad Neurol 2014;17:231-3

Introduction

Neurosyphilis (NS) is defined as an infection of the central nervous system caused by *Treponema pallidum*. Up to 4-10% of patients with untreated syphilis may develop NS.[1] These days NS rarely presents in its classical forms, tabes dorsalis or general paresis, and often presents with atypical features.[2] The clinical and laboratory features of NS have been classified into six categories.[3] Spinal syphilis was classified by Adams and Meritt into five categories.[4] Erb's paraplegia refers to patients of syphilitic meningomyelitis with very slow progression over many years and predominantly motor signs.[5] As *Treponema pallidum* can affect any part of the neuraxis, overlap syndrome affecting more than one part of the central nervous system is possible. Primary optic atrophy occurs more than twice as frequently in tabes dorsalis as in other types of neurosyphillis.[6] The overlap syndrome of Erb's paraplegia with primary optic atrophy in NS has rarely been reported in literature.

Access this article online	
Quick Response Code:	Website: www.annalsofian.org
	DOI: 10.4103/0972-2327.132648

Case Report

A 32-year-old male patient, taxi driver, presented with 4-year history of slowly progressive weakness of both lower limbs. Over next few months he developed erectile dysfunction, which was followed by bladder involvement in the form of urgency and urge incontinence. He had stiffness of both lower limbs. There was no history of numbness, imbalance while walking, and pain or paresthesias in any limb. Since last 2 months he developed a subacute increase in his weakness in form that he had to take the support of a stick to walk. He had history of promiscuous sexual behavior over the last decade. He had history of penile ulcer 5 years back which had healed over 15 days. On examination, a small scar was present on the glans penis. Central nervous system (CNS) examination revealed spastic paraplegia with 3/5 power at all joints in lower limbs. The deep tendon reflexes were all brisk with ankle clonus and plantars were both extensors. Sensory examination including joint position and vibration was normal. Rhonberg's sign was absent. There was a relative afferent papillary defect (RAPD) in the right eye, and vision was 6/24 in the right eye and 6/6 in the left eye. The accommodation reflex was normal in the right eye. Color vision was normal in both eyes. Fundus examination revealed pallor of the optic disc in the right eye

and the disc appeared chalky white on the right side, which was suggestive of primary optic atrophy [Figure 1]. The disc on the left side was normal. Magnetic resonance imaging (MRI) brain and whole spine was normal [Figure 2]. The routine blood investigations including complete blood count, random blood sugar, liver function tests, and renal function tests were normal. Erythrocyte sedimentation rate (ESR) was 20 mm/h. Chest X-ray and electrocardiogram (ECG) were normal. C-reactive protein (CRP), antinuclear antibody (ANA), and double stranded deoxyribonucleic acid (dsDNA) were normal. Vitamin B12 levels were 1,082 pg/ml. Serum copper was 110.0 μg/dl (normal 80-140 μg/dl). Human immunodeficiency virus (HIV) was nonreactive. Serum electrolytes and thyroid profile was normal. Serum Venereal Disease Research Laboratory (VDRL) was reactive. Cerebrospinal fluid (CSF) showed total cells-6/mm^3 (all lymphocytes), sugar-24 mg% (blood sugar-120 mg%), proteins-220 mg%, and CSF VDRL was reactive. CSF gram stain and acid-fast Bacilli (AFB) stain were negative. On the basis of serum VDRL and CSF VDRL being reactive the diagnosis of symptomatic NS was made. [Table 1][7] Patient had slowly progressive motor predominant myelopathy (Erb's paraplegia) with primary optic atrophy, thus having category 3 (ocular) and 4 (myelopathy) of clinical categories of NS. [Table 2].[3] Patient was started on aqueous penicillin G (24 mU/day) intravenous given in divided doses 4 hourly for 2 weeks after sensitivity testing. The plan was to follow-up the patient after 6 months for clinical and repeat CSF examination to look for clinical recovery and decrease in pleocytosis and decrease in VDRL titers.

Discussion

Non-tabetic syphilis affecting the spinal cord is rare and only sporadic case reports have been published. In 1944 Adams and Merritt classified spinal syphilis into:

1. Syphilitic meningomyelitis,
2. Spinal vascular syphilis,
3. Syphilitic spinal pachymeningitis comprising spinal cord gummata and hypertrophic pachymeningitis,
4. Syphilitic poliomyelitis, and
5. Spinal cord compression due to vertebral gumma or aortic aneurysm.[4]

Meningomyelitis comprised 15 of their 31 patients and consisted of subacute or chronic progressive paraparesis associated with variable sensory and spinchter disturbances. The variant of meningomyelitis known as Erb's paraplegia refers to patients with very slow progression over many years and predominantly motor signs.[5] In a series of nine patients of syphilitic myelopathy by Silber[8] only one patient had Erb's paraplegia. In a series of 16 patients of NS from northeast India,[9] five patients had myelopathy. Amongst them two patients had acute transverse myelitis and chronic myelopathy was present in three patients. But none of these five patients had Erb's paraplegia nor did they have optic neuropathy.

Although NS has been divided five major categories, that is, asymptomatic, meningeal, meningovascular, parenchymatous, and gummatous, these entities represent a continuum and

Table 1: Diagnostic criteria of neurosyphilis

Definitive diagnosis (requires 1 or 2 and 3)
Identification of *T. pallidum* in CSF or CNS tissue by microscopic examination or animal inoculation or by PCR
A reactive serum treponemal test
A reactive VDRL-CSF test on spinal fluid sample.
Presumptive diagnosis (requires 1 and 2 or 3):
A reactive serum treponemal test
Clinical signs of neurosyphilis
Elevated CSF protein or leucocyte count in the absence of other known causes

CSF = Cerebrospinal fluid, CNS = Central nervous system, PCR = Polymerase chain reaction, VDRL = Venereal disease research laboratory

Table 2: Diagnostic categories of neurosyphilis

Category 1 – Neuropsychiatric disorders including psychosis, delirium, and dementia
Category 2 – Cerebrovascular accident (CVA): Acute, focal neurological deficit, compatible with a CVA or radiological evidence of stoke
Category 3 – Ocular: Presentation with uveitis, visual loss, or optic nerve dysfunction
Category 4 – Myelopathy: Acute, subacute, or chronic dysfunction of the spinal cord, including tabes dorsalis
Category 5 – Seizure: Presentation with partial seizures, with or without secondary generalization, or myoclonus
Category 6 – Brain stem/cranial nerves: Signs restricted to brain stem and cranial nerves

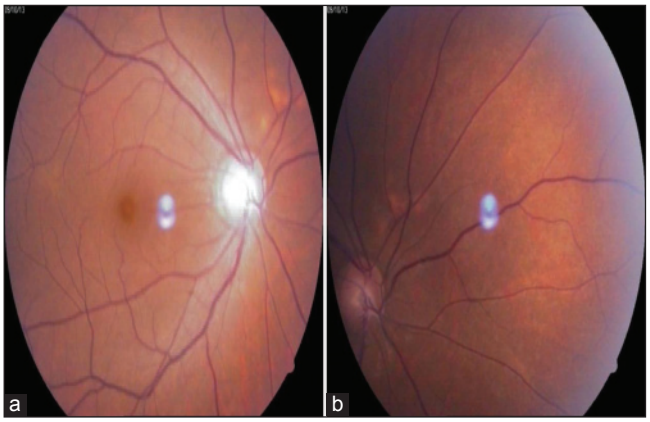

Figure 1: (a) Fundus showing primary optic atrophy (right eye). (b) Normal fundus (left eye)

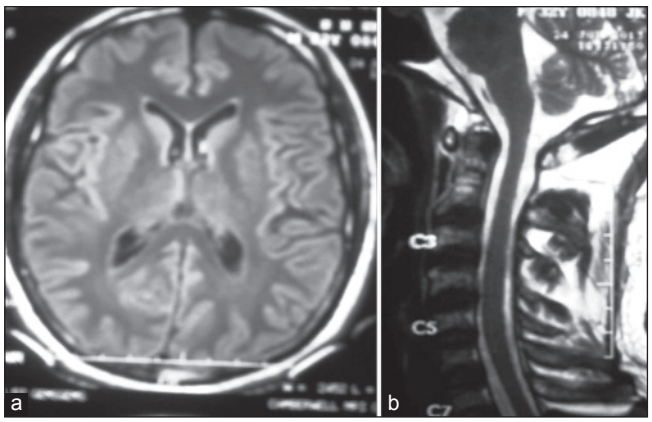

Figure 2: (a) Magnetic resonance imaging (MRI) brain-normal. (b) MRI cervical cord-normal

frequently overlap.[3] Overlap between different forms of NS is largely restricted to seizure, stroke, or neuropsychiatric features. Our patient had spinal cord involvement in the form of Erb's paraplegia along with optic nerve involvement in form of primary optic neuropathy. In the series from northeast India[9] none of the 16 patients of NS had optic nerve involvement.

Primary optic atrophy in NS begins in the intracranial portion of the optic nerve distal to the chiasma.[6] The degeneration usually first occurs in the marginal fibers of the nerve and there is thickening and perivascular round cell infiltration of the overlying membranes, especially the pia and of the connective tissue septa of the nerve. Primary optic atrophy occurs more than twice as frequently in tabes dorsalis as in other types of NS. Many patients show primary optic atrophy and pupillary changes as the only clinical manifestations of NS (so-called "preataxic" optic atrophy). Our patient showed the presence of primary optic atrophy along with presence of non-tabetic cord involvement.

About 30% of untreated individuals develop late CNS disease. The incidence of NS in HIV infected patients in between 9 and 23.5%.[10]

In the study by Timmermans and Carr,[3] out of a total of 102 confirmed patients of NS, only six patients were tested positive for HIV (5.88%). In a study by Sethi *et al*.,[11] out of 25 confirmed cases of NS, two patients were tested positive for HIV (8%). In the same study optic atrophy was present in 12% patients of NS, while none had spinal cord involvement.

Rompalo *et al*.,[12] report that the only significant difference among HIV-infected and uninfected persons suffering from syphilis was an increase in median number of genital ulcers.

The combination of noncompressive myelopathy with primary optic atrophy lands us in a diagnostic dilemma. We had to rule out other possible causes of this association like primary progressive multiple sclerosis, neuromyelitis optica, HIV myelopathy, and myelopathy associated with vitamin B12 and copper deficiency. Serum VDRL screening has started to become obsolete in workup of predominantly motor form of noncompressive myelopathy as NS affecting spinal cord has been traditionally thought to present as tabes dorsalis. Our case report highlights the fact to incorporate serum and CSF VDRL screening in workup of noncompressive myelopathy.

NS being a potentially treatable disorder, a high index of suspicion especially in patients with promiscuous sexual behavior, can lead to an accurate diagnosis even when NS presents with rare combinations of clinical involvement.

References

1. Conde-Sendma MA, Amela-Perisa R, Aladro-Benito Y, Maroto AA. Current clinical spectrum of neurosyphilis in immunocompetent patients. Eur Neurol 2004;52:29-35.
2. Madhusudhan M. Neurosyphilis. Neurol india 2009;57:233-4.
3. Timmermans M, Carr J. Neurosyphilis in the modern era. J Neurol Neurosurg Psychiatry 2004;75:1727-30.
4. Adams RD, Merritt HH. Meningeal and vascular syphilis of the spinal cord. Med (Baltimore) 1944;23:181-214.
5. Wilson SA. Neurology. Ch. 17, 2nd ed. London: Butterworths; 1954.
6. Hahn RD. Tabes dorsalis with special reference to primary optic atrophy. Br J Vener Dis 1957;33:139-48.
7. Pope V, Steiner BM. Syphilis. In: Hiseox JA, editor. Topley and Wilson's Microbiology and Microbial Infection, 10th ed. New York: Wiley and Blackwell; 2007. p. 1838-64.
8. Silber MH. Syphilitic myelopathy. Genitourin Med 1989;65:338-41.
9. Kayal AK, Goswami M, Das M, Paul B. Clinical spectrum of neurosyphilis in North East India. Neurol India 2011;59:344-50.
10. Adriana M, Allison PB, Marcus Vde O, Pedro H de CA, Melina MB, Anna MB. *et al*. Neurosyphilis and clinical variants. Rev Neurosci 2012;20:546-51.
11. Sethi S, Das A, Kakkar N, Banga SS, Prabhakar S, Sharma M, *et al*. Neurosyphilis in a tertiary care hospital in North India. Indian J Med Res 2005;122:249-53.
12. Rompalo AM, Joesoef MR, O'Donnell JA, Augenbraun M, Brady W, Radolf JD, *et al*. Syphilis and HIV Study Group. Clinical manifestations of early syphilis by HIV status and gender: Results of the syphilis and HIV study. Sex Transm Dis 2001;28:158-65.

How to cite this article: Sharma C, Nath K, Kumawat BL, Khandelwal D, Jain D. Erb's paraplegia with primary optic atrophy: Unusual presentation of neurosyphilis: Case report and review of literature. Ann Indian Acad Neurol 2014;17:231-3.
Received: 16-10-13, **Revised:** 07-12-13, **Accepted:** 12-02-14

Source of Support: Nil, **Conflict of Interest:** None declared.