CASE REPORT

YOUNG PATIENT PRESENTED WITH IMBALANCE DIAGNOSED AS A NEUROSYPHILIS

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ABSTRACT

A 30 year old female patient was presented with a history of weakness of both lower Limbs, sense of imbalance while standing and difficulty in walking with no significant past history. Upon investigation for paraperesis, the patient found to have a positive serology for syphilis. The possibility of neurosyphilis was considered, which was later confirmed by positive serology for cerebrospinal fluid and diagnosed as tabes dorsalis. The case is lengthly discussed in this report to emphasize the presence of syphilis in 21st century and for awareness of florid manifestations of tabes dorsalis for early diagnosis and proper treatment of this disabling condition.

Keywords: Neurosyphilis, Tabes dorsalis, Venereal Diseases Research Laboratory (VDRL).

INTRODUCTION

The name Syphilis is derived from the Greek word "Syphilos" meaning crippled or maimed. Syphilis is usually a sexually transmitted infection characterized by episodes of active disease interrupted by latency periods. There are three characteristic stages of Syphilis namely; Primary, Secondary and Tertiary. The tertiary stage occurs in around one third of cases and is manifested by progressive musculoskeletal lesions, aortic involvement or symptomatic central nervous system disease. The causative organism for Syphilis is a spirochete named; Treponema Pallidum which is a helically coiled organism measuring some 5-20 micrometers (μm) in length. Syphilitic infection of the nervous system results in one of the most chronic and insidious meningeal processes known. Central nervous system invasion occurs early in the course of untreated syphilis.[1] Therefore, patients not treated for persistent cerebrospinal fluid abnormalities are at risk of developing clinically apparent diseases. [2]

CASE

Clinical Presentation: A 30 year old Hindu female patient with no significant past history, admitted with weakness of both lower limbs, sense of imbalance and difficulty in walking since 4 months. Detailed history revealed of history of pain tingling and numbness over both lower limb, inability to sense pain and temperature since 6 to 8 months with no history of genital ulceration or urinary incontinence. Patient is taking mixed diet, having three children.

On examination, there was no trophic ulcer, or hypopigmented anaesthetic patch or thickened peripheral nerve trunks. The central nervous system (CNS) examination revealed normal higher mental functions and cranial nerves. The muscle of both lower limbs showed wasting and weakness (grade IV). The deep tendon reflexes (knee, ankle) and superficial reflexes (abdominal) were absent with bilateral flexor planter response. The temperature, pain and touch sensations were impaired below T6 level. The vibration and joint pain position sense were lost below knee on both sides. The Romberg sign was positive and the gait was ataxic and broad based. Ophthalmological examination revealed normal light and accommodation reflexes and so were the conjunctival and corneal reflexes. The fundoscopy examination was normal.

Investigation and Diagnosis: The complete hemogram showed normochomic normocytic anaemia with normal ESR (8 mm after 1 hr). The liver function tests (LFT), renal function test (RFT) and random blood sugar (RBS) levels were within normal limits. Vitamin B_{12} level was normal. Serum HIV was non–reactive. Radiography of brain and spine were normal. Nerve conduction velocity (NCV) and Electromyography (EMG) were normal. So serum VDRL was advised and it was positive. Treponema Pallidum Haemagglutination test (TPHA) was positive (1:160). The cerebrospinal fluid (CSF) analysis showed clear fluid with no cells with protein 8 mg % and sugar 64 mg % and positive VDRL.

Treatment: Patient was treated with injection aqueous crystalline penicillin 3 million units intravenuously 4 hourly for 21 days.

Outcome: Patient improved over one month duration in form of decreased ataxia, can able to walk without support with no broad based gait.

DISCUSSION

Tertiary syphilis has become a rarity in the 21st century. However, after a decade of decline, incidences of syphilis have increased significantly. The pathology of tertiary syphilis involving the central nervous system in obiliterative small vessel endarteritis which usually involves the vasovasorum. The clinical spectrum of tertiary syphilis includes the neuro syphilis, cardiovascular syphilis and the late gummatous syphilis.[2] Quaternary syphilis, characterized by fulminant anergic necrotizing encephalitis is seen mostly in patients with coexistent retroviral infections. A patient with neurosyphilis may be totally asymptomatic or present an acute syphilitic meningitis, meningovascular syphilis, tabes dorsalis, optic atrophy or generalised paresis of insanity. Asymptomatic neurosyphilis is characterized by reactive serology in the cerebrospinal fluid along with other features such as elevated protein and pleocytosis.[3] Acute syphilitic meningitis usually presents features of meningeal irritation. Fever is rarely observed. Involvement of cranial nerves especially of 7th, 8th, 6th and 2nd in order of decreasing frequency is common. Meningitis can be self limiting but can present more severe in later forms. The most common clinical syndrome of meningovascular syphilis is stroke; in a relatively young adult it involves the middle cerebral or basilar arteries.

Tabes dorsalis is a slowly progressive disorder involving the posterior columns and posterior roots of the spinal cord. Unlike in syphilitic meningitis and general peresis, where Treponema pallidum (T. pallidum) could be demonstated early in CSF, in tabes dorsalis T. Pallidum cannot be demonstrated in CSF or other tissues, suggesting an immunological attack on spinal cord[9]. With the exception of tabes dorsalis, the clinical syndrome is rarely so clear cut, as to permit the diagnosis of neurosyphilis in face of negative serological findings[10]. A clinical diagnosis of tabes dorsalis is more likely in patient lightening pains, ataxia, absent tendon reflexes, Argyl-Robertson pupil and positive Romberg sign. In atypical cases, only the results of serological testing and CSF examination may lead to the correct diagnosis. Other features of tabes dorsalis are bowel and bladder disturbances, gastric crisis, impotence, visual failure, papillary abnormalities, Charcot's arthropathy and perforating ulcers[11].Usually sensory symptoms especially, pain,precede ataxia. This preataxic stage lasts for 2 to 5 years. The rate of progression of disease can be roughly assessed from the duration of the pre ataxic stage. Longer the pre ataxic stage, slower will be the subsequent deterioration[12] .

The diagnosis of Syphilis is performed by serological and non serological tests. The serological tests can be further sub classified into treponemal and nontreponemal tests. The non serological tests include; dark filed microscopy, direct immunoflouroscence and demonstration of Treponema Pallidum in tissues.[6] Current recommendations for evaluation of neurosyphilis in any patient with positive serology for syphilis include those with focal neurological signs, patients with other late syphilis and cases of suspected treatment failure.

Our patient presented with characteristic features of tabes dorsalis except gastric crisis, impotence,visual disturbance, Argyl-Robertson pupils, and Charcot's arthropathy. In the early stages, it is commoner to find that the light reaction is present but reduced in amplitude. Completed Argyl-Robertson pupil appears late in the disease.Though positive serum VDRL often is a clue to presence of neurosyphilis[7],reactive VDRL in the spinal fluid is a reliable evidence of past or present neurosyphilis. Normal LFT, RFT, RBS, radiography of the spine and brain, normocytic normochromic anemia and absence of hypopigmented anesthetic patches or enlarged peripheral nerve trunks, in the present case, exclude other possible differential diagnosis like, alcoholic neuropathy, diabetes mellitus, spinal trauma, syringomyelia,vitamine B_{12} deficiency and leprosy respectively.

In the preantibiotic era,tabes dorsalis accounted for about 1/3rd of patients with neurosyphilis[3]. In recent years, late syphilis has become rare in most of the STD clinics in India. However, tabes dorsalis has been reported in association with cardiovascular syphilis or general paresis. Tabes dorsalis has also been reported to occurring following standard treatment for secondary syphilis. This may be due to the apparent persistence of T. Pallidum in humans and animals even after standard penicillin therapy for early acquired syphilis[5].

With a rise in the prevalence of HIV infection, clinicians will come across patients presenting with more florid manifestations of syphilis. Neurosyphilis may present without neurological symptoms in half of HIV infected patients with serological evidence of syphilis[5]. There are several reports to show that patients with HIV are more likely to progress to neurosyphilis in the first two years after diagnosis, despite standard treatment. Therefore, in all patients co-infected with syphilis and HIV, the CSF analysis should be done for evidence of neurosyphilis[15] and treated accordingly with high dose intravenous penicillin[5].

Penicillin has remained the drug of choice for all stages of Syphilis.[8] A single dose of Benzathine penicillin in a dose of 2.4 million units resulted in cure for more than 95% cases of primary Syphilis.[9] Late latent syphilis and late syphilis without evidence of involvement of central nervous system has to be treated with 3 weekly doses of 2.4 million units of Benzathine penicillin. In contrast, neurosyphilis requires treatment with crystalline penicillin with a dosage regimen of 2 to 6 million units every 4 hours for a total of 10 to 14 days. An alternative is to use aqueous Penicillin G procaine at a daily dosage of 2.4 million units intramuscularly, along with 500 mg of oral Probenecid every 6 hours for a course of 14 days.[10] Recommendations for treatment remain the same for HIV coinfected patients with syphilis. There is no data supporting the use of other antibiotics for the treatment of neurosyphilis, however the use of certain 3rd generation Cephalosporins and Azithromycin may merit further evaluation. In a recent study by Bai et al, published in the international journal of STD/AIDS, Azithromy-

cin achieved a higher cure rate than Benzathine penicillin for early syphilis.[11]

CONCLUSION

Though antibiotics cannot reverse the extensive changes, they may arrest the progression of tabes dorsalis in 50 per cent of cases. Therefore, in the era of HIV infection, for the early diagnosis and prompt treatment of tabes dorsalis, alertness to the possibility and awareness of its pleomorphic clinical and laboratory manifestations is crucial.

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