

ERB'S SYPHILITIC PARAPLEGIA (A case report)

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Summary

A 45 years old married lady was admitted with spastic paraplegia of insidious onset without any sensory deficit. Serological studies of blood and CSF for syphilis were positive and cerebrospinal fluid showed increased cells and protein content. She responded well to parenteral penicillin therapy, showed gradual subject improvement and appreciable decrease in cellular and protein content of the CSF. Her husband also was found to have latent syphilitic infection. Review of relevant literature showed no report of such paraplegia following syphilis in recent times. The case is being reported with a short discussion on the clinical features of neurosyphilis.

Incidence of venereal diseases including syphilis is on the increase especially among the younger age groups^{1,2}. However, its complications are scarce because of better awareness and understanding among masses resulting in early diagnosis and adequate treatment. In comparison to prepenicillin era, neurosyphilis is rare these days. A case of Erb's syphilitic paraplegia, a rare variety of neurosyphilis who improved with penicillin therapy under our care is being reported.

Case Report

A 45 years old married lady was admitted with complaints of gradually deteriorating weakness of the lower limbs of one month's duration leading to bedridden stage a week before admission. She also had incontinence

of urine of 15 days' duration. There was no history of preceding febrile illness or of fall or injury to the spine. There was no sensory disturbances or deficit in the beginning or at any stage of illness. Past, personal and family histories were non-contributory. She was married for 27 years and had three children. No past history of abortion or miscarriage. The patient's husband was also examined in detail but there was no clinical abnormality.

On examination, she was found to be moderately built and nourished. Pulse-72/mt. regular. Temperature was normal. BP was 130/84 mmHg. There was no pallor, icterus, lymphadenopathy or pedal oedema. No skin rashes, eruptions or subcutaneous nodules were seen. Heart, lungs and abdomen were normal. Central nervous system showed the following: the lady was fully conscious, well oriented and had normal unimpaired memory and speech. Ocular fundi were normal. Pupils were normal, circular, equal on both sides with light and accommodation reflexes normal. There was no

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cranial nerve abnormality. Motor and sensory systems of upper limbs were normal. Lower limbs showed increased (spastic) tone and loss of power (Grade I-II/IV) in all muscle groups. Deep reflexes (knees and ankles) were exaggerated. Ankle clonus was present. Plantar showed extensor response on both sides. There was no sensory loss or deficit of any modality and no band of anaesthesia could be detected on the trunk. Bladder was incontinent.

Investigations showed Hb — 12.5 gms%. TLC — 7,500/cu. mm DLC - P 57%, E. 4%, L. 38%, M. 1%. Urine and stool were normal. Urine Culture was sterile. Blood Urea was 30 mgm%, Blood sugar Fasting 86 mgm%, PP-96 mgm%. Serum cholesterol was 210 mgm% X-Ray Chest and ECG were normal. X-Rays of the skull and dorsolumbar spine did not show any abnormality. Blood WR was reactive Kahn was positive (32 KT) and VDRL reactive in 16 DIL. CSF showed normal pressure, cells were 40/cu.mm (mostly lymphocytes), protein 170 mgm%, globulin was increased, sugar and chlorides normal. CSF WR, Kahn and VDRL were positive. The husband's blood WR Kahn and VDRL, were positive but all CSF results were negative.

The history of insidious onset with evidence of spastic paraplegia without sensory loss, combined with positive blood and CSF serology for syphilis, led to the diagnosis of Erb's paraplegia. Patient was treated with PAM 600 mg once a day for 20 days along with physiotherapy and other supportive treatment. She showed significant improvement within two months of treatment. She is now able to move the limbs but not able to walk without support. The repeat blood and CSF serology after two months are still reactive for syphilis, but the cell (2/cu mm) and protein (80 mg%) con-

ent of CSF have shown appreciable decrease. She is under follow up and continues to have physiotherapy along with other symptomatic treatment.

Discussion

Though considerable knowledge about the clinical aspects and pathology of syphilis was acquired between the sixteenth and the nineteenth centuries, detailed descriptions of involvement of central nervous system were available only in the last century.

Spirochaetes are known to invade the central nervous system even in the primary stage, though symptomatology may appear later. CSF abnormalities have been found by Nicolou at this stage showing a mild lymphocytosis with positive serological test in 1.5% of cases³. In the secondary stage CSF abnormalities are seen in 36% to 80% of cases with 6% cases showing positive serological test in cerebrospinal fluid³. There may not be any serious sign or symptom except occasional headache. Very occasionally acute and fatal meningo-encephalitis may be seen in secondary syphilis. The so called 'neuro-relapse' also occurs at this stage due to incomplete treatment and manifests with convulsions, coma, ocular palsies, aphasia and hemiplegia.

The central nervous system involvement in tertiary syphilis occurs in atleast 10% of infected individuals. These are divided into two groups: the meningo-vascular syphilis and the parenchymatous syphilis. Parenchymatous syphilis occurs late (10-20 years after infection) and is described as general paresis of insane and tabes dorsalis. Meningo-vascular syphilis of the brain is described as cranial pachymeningitis, gummatous leptomeningitis, cerebral endarteritis and gumma. Spinal involvement in meningovascular syphilis are cervical pachymeningitis, meningomyelitis, spinal endarteritis, radiculitis, and Erb's syphilitic spinal paralysis.

The clinical presentation of spinal syphilis is usually paraplegia, except in radiculitis where root pain of the involved nerves may be the only symptom. Cervical pachymeningitis produces inflammation of the dura-mater with subsequent thickening and adhesion to arachnoid and pia-maters. Thus the nerve roots are cuffed and obstructed producing pain and are ultimately destroyed producing atrophy of the muscles and paraplegia with sensory loss. A quadriplegia may result if the upper segments of the cervical cord are involved. Similarly meningomyelitis produces inflammation of the meninges as well as softening of the grey matter of the spinal cord—resulting in tranverse lesion of the spinal cord. These lesions usually are chronic, but thrombosis of a vessel supplying the spinal cord may result in acute paraplegia. The involvement of the spinal cord is limited only to a few segments—resulting in lower motor neurone paralysis below the site of lesion.

Erb described an uncommon spinal cord lesion seen especially in middle aged persons⁴. The clinical picture is a slowly progressive spastic paraplegia without any sensory involvement or with minimal sensory loss. The bladder is involved early producing urinary incontinence. The incidence of Erb's paraplegia appears to be low, as recent large reviews of non-traumatic paraplegias do not include a single case of paraplegia due to syphilis^{6,7}.

The line of management of the syphilitic paraplegia, indeed of neurosyphilis, used to vary from country to country—in the pre-penicillin days. However, the standard line of treatment now is parenteral penicillin injections in addition to other supportive therapy. A daily dose of 600 mg of PAM given intramuscularly for 20 days is the most accepted line of treatment. Erythromycin 500 mg 6 hourly for 15 days is used as an alternative line of therapy in case of sensitivity to penicillin⁵.

The initial assessment of blood and CSF serology as well as CSF biochemistry must be done prior to initiation of treatment. These investigations should be repeated atleast at six monthly intervals and further course of penicillin injection be instituted if either clinical or biochemical improvement is not adequate.

Treatment with bismuth, iodides, arsenobenzenes and pyrexial doses of E. Coli and malarial parasite are largely outmoded forms of treatment now and resorted to only in an exceptional case.

Care of urinary bladder to prevent infection and further complications thereof is the hall mark of supportive treatment in paraplegics. The involvement of bladder in the paraplegics with the resultant urinary tract infection, ascending pyelonephritis and renal failure is usually the dominant complicating feature of the illness and largely influences the prognosis^{6,7}.

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