

# General paresis of the insane - A rare case report

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**Abstract** General Paresis of the Insane (GPI) is a meningoencephalitis as a result of direct invasion of *Treponema pallidum* into the cerebrum, typically develops between 15 and 20 years after initial infection. Mr. X is a 50 year old painter referred to Venereology Department from Neurology Department with 3 years history of gradually progressive behavioural disturbances along with disturbances in speech & difficulty in writing. Blood VDRL & CSF VDRL was found to be reactive. A diagnosis of GPI was made and the patient was treated appropriately. He showed remarkable improvement following treatment.

**Key words**

General paresis of insane, crystalline penicillin, lumbar puncture.

## Introduction

General Paresis of the Insane also known as Dementia Paralytica, Paretic Neurosyphilis presents as combination of symptoms that can mimic most neurological & psychiatric symptoms. Syphilitic infection of the nervous system has shown a tremendous decline in this century with an attenuated & atypical presentation which leads to the diagnostic problem.<sup>1</sup> Remarkable improvement following treatment in a case of neurosyphilis is very rare. We report a rare case of GPI who showed marked improvement following treatment.

## Case report

Mr. X is a 50 year old painter referred from Neurology Department to rule out neurosyphilis with 3 years history of gradually progressive

behavioral disturbances characterized by irritability, social withdrawal, sleep disturbances, sudden crying spells & anger bursts. He also had forgetfulness, difficulty in remembering his name, inability to find his way back home and unable to recognize his family members. He was first treated in psychiatry department both as outpatient & inpatient but there was no improvement. Later seen in neurology department where he was diagnosed as a case of pre-senile dementia. Finally he was referred to Venereology department to rule out neurosyphilis. There was no history of any treatment taken for genital ulcer, skin rashes etc. History of high risk behavior could not be elicited satisfactorily.

Mental state examination revealed labile affect, impairment in orientation, recent memory, intellectual functioning, judgement & insight. His speech was monotonous without any modulation with grammatical errors & faulty enunciation. His handwriting was grossly impaired, illegible with missed letters and transposition of letters (**Figure 1**). There was increase in deep tendon reflexes & bilateral positive Hoffman & palmomental reflexes. Gait

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GPI, progress toward the general acceptance by the medical community of this idea was only accomplished later by the eminent 19<sup>th</sup> century syphilographer Alfred Fournier. In 1913, all doubt about the syphilitic nature of paresis was finally eliminated when Noguchi & Moore demonstrated the spirochaetes in the brains of paretics.

The diagnosis of GPI is most often missed because of the insidious behavioral changes that precede the onset of actual clinical presentation. Our case also presented with behavioral abnormality much before the onset of other clinical signs. However the feature of dementia which was present in our patient was a common presentation of GPI.<sup>2</sup> Psychiatric manifestations of GPI includes memory loss both recent & remote, personality changes, neurosis, euphoria, overactivity, depression, reduction in mental capacity, disorientation, failures of judgement in an otherwise astute person. Grandiose delusions & megalomania, although dramatic manifestations, occur only in 10-20% of cases.<sup>3</sup>

In addition to psychiatric symptoms, neurologic symptoms are also common in GPI which includes dysarthria, Argyll Robertson pupil, flattening of facial lines, tremors of tongue on protrusion (tongue tremor), abnormality of deep tendon reflexes, speech abnormalities (dysnomia, global aphasia etc), impaired handwriting, hypotonia, cranial nerve palsies & sphincter incontinence. Adult onset seizures or ischemic attacks without the presence of psychiatric symptoms may be the initial presentation in 15-20% of patients with paresis. This is especially prevalent in those cases of paresis in which the onset of symptoms is sudden and not gradual.<sup>3</sup> The combination of hemiparesis, convulsions & aphasia has been termed as **Lissauer's** type of GPI.<sup>4</sup> The common triad of pupillary abnormality, tremors & dysarthria was not evident in our case and only

one component of the triad i.e. dysarthria was present. This isolated clinical feature by itself is not sufficient for a diagnosis of GPI but when present in conjunction with dementia and positive VDRL in serum and/or CSF, it has a diagnostic specificity.

The diagnosis of neurosyphilis depends upon reactivity of serologic tests which currently comprise of a screening test, a confirmatory test and CSF examination. Commonly used screening tests include VDRL and RPR. If non-treponemal screening test is reactive, it is usually confirmed by more specific treponemal tests like FTA-Abs and TPHA test. Standard parameters of neurosyphilis on CSF examination include mononuclear cell count greater than 5 to 10 cells/mm<sup>3</sup>, protein concentration greater than 40mg/dl and a reactive CSF VDRL.<sup>5</sup> CT-Brain shows decreased attenuation in the cerebral white matter of frontal & parietal lobes, with enlarged cortical sulci & ventricular dilation. Such changes are similar to those seen in demyelinating disorders.<sup>6</sup>

Penicillin remains the mainstay of treatment for neurosyphilis. The CDC recommends that neurosyphilis be treated with intravenous aqueous crystalline penicillin G, 18 to 24 million units, divided into six doses or as a continuous infusion for 10 to 14 days.<sup>7</sup> An alternative regimen is intramuscular procaine penicillin G, 2.4 million units once daily with oral probenecid, 500mg four times daily, both for 10 to 14 days. For penicillin-allergic patients, intramuscular or intravenous ceftriaxone, 2g once daily is recommended as an alternative.<sup>8</sup> Penicillin skin testing and desensitization are reserved for those individuals in whom the safety of ceftriaxone is of concern. Azithromycin could potentially be used to treat neurosyphilis because of its excellent CNS penetration.<sup>9</sup>

Neurosyphilis among elderly patients poses lot of problems. New cases are not adequately detected and delay in diagnosis is common; often clinical presentation does not follow the traditional course; mental changes may imitate other types of dementia and psychiatric disorders. All elderly patients with neurological or psychiatric disorders of doubtful etiology should have syphilis serology checked routinely.

The fact that neurosyphilis can mimic any psychiatric & neurological condition, is exemplified by the fact that our patient was treated by the Psychiatrist then by the Neurologist, finally he landed in Venereology Department. Nowadays Neurosyphilis become very rare and patient has shown marked improvement following treatment which is an interesting feature in this case.

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