General Paresis of Insane: A Forgotten Entity
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Abstract: The manifestations of CNS syphilis are unfamiliar to a differential of patients with dementia to many physicians today as of the relative rarity of this condition. This is a classical case report of a patient with syphilis and dementia in a 55 old female. General paresis of insane is a progressive disease of brain leading to mental and physical worsening. It is important to consider tertiary syphilis in the differential diagnosis of dementia. Conventional presentations of neurosyphilis such as tabes dorsalis and general paresis of insane are read in textbooks only and rarely encountered in clinical practice in the 21st century.

Keywords: Dementia, general paresis, neurosyphilis, psychiatric manifestations

INTRODUCTION
Syphilis is believed to be brought to India by the Portuguese in the 16th century [1]. Syphilis has expanded rapidly in past two decades. The increase started in the late 1970s, due to the alteration in sexual behavior [2]. Neurosyphilis is a mysterious disease because it imitates psychiatric diseases leading difficulties in differential diagnosis. In addition, antibiotics may change its natural course, and therefore, its clinical manifestations [3]. Neurosyphilis is rather an unusual cause of dementia characterize by a rapidly progressive course and psychiatry symptoms [4].

Paretic neurosyphilis or general pareses usually develop 15-20 years after infection. PARESIS is an acronym ((involvement of Personality, Affect, Reflexes, Eye, Sensorium, Intellect and Speech). Upon clinical suspicion, diagnosis of neurosyphilis is confirmed by a reactive cerebrospinal fluid (CSF) VDRL, Treponem pallidum Hemagglutination assay (TPHA). CSF examination was mandatory in neurosyphilis diagnosis [4].

Asymptomatic cases and cases with ill-defined syndrome become more common than the classic presentation of tabes dorsalis and general paresis. In this article, we present a case of neurosyphilis with progressive cognitive changes and intractable behavior and psychiatric problems whose primary and secondary phases were not detected.

CASE REPORT
We present a case of a 55 year old female presented with complaints of forgetfulness, irritability, crying spells, aggressive behavior, hallucinations and illusions over the last 5-6 months. Her symptoms had started with features of withdrawn behavior, became less communicative than before. She was quite most of the time and she was not oriented to time, place and person. She can not recognize the immediate family member, as well as she cannot engage in long conversations like before. The patient had problem of forgetfulness, she often would venture out and forget the way back. She had started remaining fearful and would prefer to stay indoors and keep the doors and windows of the house closed. She would often burst into tears for no apparent reason. She was taken to psychiatrist and put on antipsychotic drugs with no benefit. Her medical and family histories were not remarkable. She did not have multiple partner sexual contact. Her folstein mini mental score was 7/30. Our patient was not fully oriented, memory assessment revealed a severe verbal learning impairment with an extremely low ability to retain new information. She also demonstrated difficulty with remembering autobiographical and personal information. A severe dysexecutive syndrome was also documented. Other observations included defects in judgement, emotional lability, delusions and inappropriate social and moral behaviour. She used to soil her clothes with urine and stool. Patient had unsteady gait disturbance. Her neurological examination showed— the right pupil was 2.5 millimeters with minimum reaction to light. The left pupil was 6 millimeter and reactive to light. Unequal pupils with one of them reactive to light: the other not reactive, pupils react normally to convergence accommodation. The rest of cranial nerves except 8th
having mild bilateral conductive deafness were intact with good gag reflex. Reflexes were 2+ all over, nystagmus absent. Babinski was positive on right side, muscle power was 3/5, unsteady gait. Laboratory workups, including a complete and differential blood count, serum electrolytes and glucose, liver and renal function tests, thyroid function tests, serum B12 and folate levels were normal. Serological tests for HIV and hepatitis B/C were negative. No autoimmune and or inflammatory markers were found. In addition, cerebrospinal fluid analysis showed pleocytosis (20 leukocytes/mm3, mainly lymphocyte), elevated protein levels and normal glucose. Serum VDRL was positive in 1:128, the serum TPHA (Treponem pallidum Hemagglutination assay) was positive at 1; 2560 dilution. CSF yielded positive for VDRL 1:4 and CSF-TPHA – 1:640. Although brain MRI was negative. EEG was within normal limits. The patient was diagnosed as having dementia due to neurosyphilis. She was started on 24 million units of aqueous penicillin G IV quid for 21 days and clonazepam 2 mg/day orally and resperidone 2mg/d, for behavior disorders and agitation. During her follow up, we did not observe any improvement in her psychiatric symptoms, cognitive functions; urinary and fecal incontinence.2.4 million units’ benzathine penicillin/month IM prophylaxis was planned for maintenance treatment. CSF was not re-evaluated.

DISSCUSSION

Syphilis is a multisystem chronic infection caused by Treponema pallidum. There is widely held clinical opinion that syphilis has disappeared. Contrary to this, there are sporadic cases being reported across the country [1]. Rapidly progressive dementia (RPD) associated with neuropsychiatric symptoms is the most common form of presentation of general paresis and includes a series of disturbances such as personality changes, amnesia, delusions, hallucinations and delirium[4].

No clinical or dermatological symptoms or sign was found related with primary and secondary stages of syphilis in our patient’s history. It is noteworthy that clinical picture emerged with tertiary syphilis first. This can be explained by the change in the natural course of the disease by widespread antibiotic use and not remembering the past of the patient and her family [5]. However, although disease related symptoms are generally observed in primary syphilis, only 1-2% of patients with secondary syphilis are asymptomatic [6]. Patients enter into latent/asymptomatic phase which only serological findings are present after recovery from secondary stage. At this stage CSF is generally normal and if there are abnormal CSF finding then it is evaluated as asymptomatic neurosyphilis. Tertiary syphilis develops in more than 1/3 of untreated patients [7]. Response to treatment is inadequate in parenchymatous neurosyphilis cases compared to syphitic meningitis and meningovascular syphilis patients. This is mainly due to irreversible neuronal damage in general paresis. The most effective treatment is high dose IV crystallized penicillin [8].

Our case had classical presentation of general paresis of insane as it typically started with affective symptoms which gradually progressed to cognitive decline leading to frank dementia. General paresis of insane is a parenchymatous neurosyphilis (dementia paralytica) which is a form of tertiary syphilis develops 10-20 years after the primary infection.

CONCLUSION

Syphilis is still prevalent, especially in particular sections of the population. Late complications can be somewhat less of an issue than the preantibiotic era, however vigilance to the probability of late neurosyphilis and appreciation of clinical manifestation of late syphilis are crucial if these form of disease are to be diagnosed and treated adequately. The main considerations must be vigilance in finding, treating, and preventing early syphilis. Since all forms of syphilis, especially advanced neurosyphilis are less common than the glory days of syphilis, it is important to educate others and to remind ourselves of the multiple faces of the great actor, lues venerea.

Conflicts of interest

The authors have no conflicts of interest to declare.

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REFERENCES