

Reversible dementia: The imitation game

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CASE STUDY

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ABSTRACT

Rapidly progressive dementia (RPD) is an emergency in behavioural or cognitive neurology. Many rare neuroinfections like Neurosyphilis may be missed, if they are not thoroughly evaluated. We report a patient with subacute onset and progressive cognitive decline, extrapyramidal involvement and myoclonic jerks who was initially suspected as probable autoimmune encephalitis or Creutzfeldt-Jakob disease (CJD). Investigations revealed positive serum and cerebrospinal fluid (CSF) Venereal Disease Research Laboratory test (VDRL). On treatment with penicillin, he developed Jarisch-Herxheimer reaction and was treated symptomatically. After two weeks of penicillin, he improved significantly and except for mild short term memory recall, he is asymptomatic for last two years.

Key Words

Neurosyphilis, VDRL, rapidly progressive dementia

Implications for Practice:

1. What is known about this subject?

Rapidly progressive dementia (RPD) is an emergency in

behavioural or cognitive neurology. Neurosyphilis is a great mimicker which can present with RPD.

2. What new information is offered in this case study?

We report a case of RPD who was initially suspected as probable autoimmune encephalitis or CJD and was later diagnosed as Neurosyphilis. During treatment he developed complications which were successfully managed.

3. What are the implications for research, policy, or practice?

This case study highlights that Neurosyphilis should be included in the evaluation of RPD.

Background

Rapidly progressive dementia (RPD) is an emergency in behavioural or cognitive neurology. Though there is no standard time bound definition for RPD, practical time used is less than six months.^{1,2} But since CJD can present as late as one year, many consider one year as the cut off period. The differential diagnosis is vast and in the modern era many infections are missed initially in the diagnostic work up of RPD.

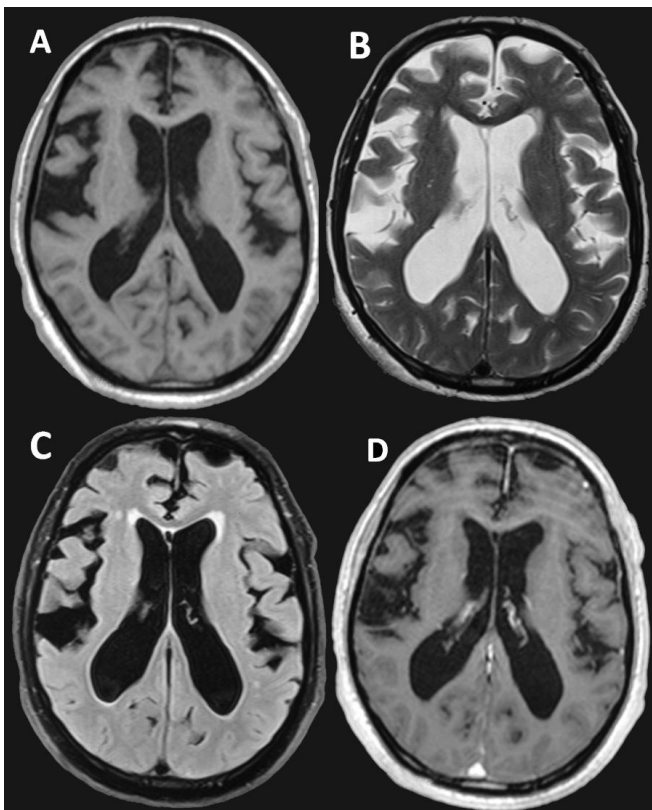
Case details

A 56-year-old man presented with progressive behavioural disturbance over five months. He had anger outbursts, aggressiveness and social disinhibition. His family noticed slurring of speech with merging of syllables. These symptoms progressed rapidly over the last two months along with urinary incontinence and myoclonic jerks. He had no history of neurologic or psychiatric disorders.

On admission, the patient was conscious and oriented but was irritable with spastic speech, impaired attention, and significantly impaired short-term recall. There were myoclonic jerks, mild rigidity and generalized hyperreflexia with release reflexes. Cranial nerves including pupils, strength, sensory and coordination were normal. The Romberg sign was absent. Medical history included

megaloblastic anemia due to vitamin B12 deficiency in 2011 which improved with treatment. An MRI showed cortical atrophy predominantly in frontal and temporal lobe with mild subcortical gliosis (Figure 1A-D). CSF analysis showed elevated protein (140mg/dL), reduced glucose (45mg/dL, blood glucose 85mg/dL) pleocytosis (120 cells, 70 per cent lymphocytes). The differential diagnoses for the patient's progressive cognitive decline, myoclonus and memory loss were autoimmune/paraneoplastic encephalitis (anti-Hu, anti-Ma, anti-NMDA, anti-GAD and anti-VGKC antibodies), chronic infections (Tuberculosis, Neurosyphilis, CJD) and neoplastic lesions.

Figure 1 A-D: Cranial MRI showing cortical atrophy predominantly in frontal and temporal lobe with mild subcortical gliosis in T1W, T2W, FLAIR and Gadolinium enhanced images



CSF Gram stain, culture, TB-PCR, HSV PCR was negative. EEG showed diffuse theta range slowing. Serum HIV, thyroid peroxidase, antinuclear antibodies, thyroid, renal, and liver function were unremarkable. CT chest, abdomen, and pelvis were normal. The treatment decision of empirical steroids was discussed with patient's family as they could not afford autoimmune/paraneoplastic panel investigations. Next day, CSF Venereal Disease Research Laboratory (VDRL) (1:32) and treponemal pallidum hemagglutination (TPHA) turned positive. The serum VDRL was also positive.

The patient received intravenous aqueous crystalline penicillin G (24 million units per day) along with olanzapine. The treatment with penicillin caused abrupt onset of fever, chills and one episode of generalized tonic clonic seizures. Considering Jarisch-Herxheimer reaction he was treated with phenytoin, aspirin and short course oral steroids (prednisolone 60mg for three days). He received penicillin for 14 days and improved significantly (MMSE 27/30) in one month with residual deficit in short term recall. His wife was also detected to be serum VDRL & TPHA positive. Her CSF VDRL was negative and was treated with three doses of Benzathine Penicillin 2.4MU IM weekly. The patient later revealed about sexual contact with several female sex workers 20 years before. But he denied any history of genital lesions. His CSF was repeated at the end of treatment which showed no cells and normal sugar and mildly raised protein (65mg/dL). CSF examination was repeated thrice in six monthly intervals each showing normal biochemistry and no cells. There was fourfold reduction in VDRL titre. Qualitative CSF VDRL is still positive after two years of follow up.

Discussion

Though there is no standard definition for RPD, one year is taken as the cut off period by most experts. Many patients labelled as RPD may not satisfy the diagnostic criteria of dementia and the term rapidly progressive cognitive decline is preferred in such scenarios.

Neurosyphilis is a great imitator and we cannot afford to miss it even now. In the US, the annual incidence of syphilis in the US decreased 36-times from a peak of 72 cases per lakh (1943) to 2.1 cases per lakh (2000). But it then increased to 4.5 cases per lakh (2008). Very few credible data on prevalence of neurosyphilis in India are available. As in our patient, many cases of neurosyphilis may be asymptomatic or having nonspecific symptoms during the stage of primary and secondary syphilis. VDRL is a simple and effective screening test for syphilis and must be included in any evaluation protocol of rapidly progressive dementia. The widely used mnemonic of VITAMINS (Vascular, Infectious, Toxic-Metabolic, Autoimmune, Metastases/Neoplastic/paraneoplastic, Iatrogenic, Neurodegenerative, Systemic/Seizures) is helpful in the evaluation of RPD.² Seven essential investigations required in all patients with RPD are Gad-enhanced MRI Brain, CSF analysis, EEG, HIV, VDRL, TFT, anti-TPO and Vitamin B12.

The parenchymal Neurosyphilis, General paresis of Insane (as in our patient) develops 15-30 years after primary infection. GPI can present with progressive dementia or

psychiatric manifestations like delusions of grandeur, hallucinations, illusions or inappropriate behavior.³ Being a great mimicker, it can have other neurological abnormalities like hyperreflexia, dysarthria, chorioretinitis, optic neuritis, and pupillary abnormalities. As in our patient, imaging shows frontal and temporal atrophy with subcortical gliosis. Imaging abnormalities of neurosyphilis are meningeal enhancement, hydrocephalus, gummas, periostitis, generalized cerebral atrophy, and stroke.⁴ Rarely it can mimic herpes encephalitis and normal pressure hydrocephalus.⁴ In the present era, neurosyphilis should be excluded in anyone suspected of having limbic encephalitis or CJD since rapidly progressive behavioural abnormalities are common feature in all these conditions.⁵

Conclusion

Neuroinfections like syphilis are still relevant in the evaluation of rapidly progressive dementia. Early diagnosis and treatment is the cornerstone for good prognosis.

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PEER REVIEW

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CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

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PATIENT CONSENT

The authors, *Vishnu VY, Modi M, Ahuja CK, Goyal MK, Lal V* declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).
3. This submission is compliant with the requirements of local research ethics committees.